RESEARCH PAPER INTERNATIONAL JOURNAL OF PURE MEDICAL RESEARCH

Cardiology

KEYWORDS: Ebstein's anomaly, Wide complex tachycardia.

WIDE COMPLEX TACHYCARDIA IN ADULT PATIENT WITH EBSTEIN'S ANOMALY: WHAT IS THE MECHANISM?

Volume-4, Issue-9, September - 2019 ISSN (O): 2618-0774 | ISSN (P): 2618-0766

Zied Frikha	University Hospital of Nancy, Department of Cardiology, Vandoeuvre-Les-Nancy, France
Mohamed El Minaoui*	UPR of Cardiology Rabat, Morocco *Corresponding Author elm.mhd@gmail.com
Olivier Huttin	University Hospital of Nancy, Department of Cardiology, Vandoeuvre-Les-Nancy, France
Jean-Marc Sellal	University Hospital of Nancy, Department of Cardiology, Vandoeuvre-Les-Nancy, France
Etienne Aliot	University Hospital of Nancy, Department of Cardiology, Vandoeuvre-Les-Nancy, France
Yves Juilliere	University Hospital of Nancy, Department of Cardiology, Vandoeuvre-Les-Nancy, France
Nicolas Sadoul	University Hospital of Nancy, Department of Cardiology, Vandoeuvre-Les-Nancy, France
Béatrice Brembilla-Perrot	University Hospital of Nancy, Department of Cardiology, Vandoeuvre-Les-Nancy, France

Article History
Received: 12.04.2019
Accepted: 03.07.2019
Published: 10.09.2019

ABSTRACT:
We present the case of a 60-year-old female with a history of Ebstein's anomaly diagnosed at childhood admitted to our hospital for wide complex tachycardia. Programmed ventricular stimulation was negative. Electrophysiology study revealed an atrial rhythm and programmed atrial stimulation induced atrial tachycardia identical to the clinical tachycardia with multiple circuits . Radiofrequency ablation was not performed because of the technical difficulties. At two months follow up, the patient's cardiac condition worsened and echocardiography revealed worsening tricuspid regurgitation. She underwent surgical repair. The postoperative course was uneventful.

1. INTRODUCTION

Ebstein's anomaly (E.A) represents less than 1% of all congenital heart disease [1]. Patients with E.A are predisposed to arrhythmias because of abnormal cardiac anatomy. We report a case of wide complex tachycardia in a patient with this anomaly.

2. CASE REPORT

A 60-year-old Caucasian female with history of E.A diagnosed at early childhood. Presented with dyspnea and palpitations. On admission, she was at NYHA functional class II. SBP of 120/80 mm Hg and heart rate of 200 beats per minute. Holosystolic murmur along the low left sterna border in auscultation. The 12-lead ECG during tachycardia disclosed a wide QRS tachycardia and a left bundle branch (LBB) morphology with late precordial transition and left axis deviation **(Figure 1)**. The tachycardia was resolved spontaneously, and the control ECG tracings suggested a sinus rhythm with right bundle branch block and no signs of ventricular preexcitation **(Figure 2)**.Transthoracic echocardiogram (TTE) demonstrated not only the presence of E.A of the tricuspid valve, with apical displacement of the septal and posterior leaflets of the valve (15mm/m2) but also the presence of severe tricuspid regurgitation (TR) and marked enlargement of the right atrium (area = 70cm2) **(Figure 3)**. The systolic function of both ventricles was preserved. An electrophysiology study was performed and revealed that the rhythm was atrial tachycardia with 2:1 anterograde atrioventricular conduction, mimicking a sinus rhythm **(Figure 4)**. Atrial stimulation stopped atrial tachycardia but several atrial tachycardias of different origins were induced by atrial stimulation. 1:1 atrial tachycardia developed with isoproterenol. Programmed atrial stimulation induced atrial tachycardia with 1:1 conduction identical to the clinical tachycardia **(Figure 5)**. Programmed ventricular stimulation performed up to 3 extrastimuli in control state and after isoproterenol was negative. After exercise test, the atrial rate increased slightly and was associated with lesser degrees of A-V block and increased ventricular rate (1:1 conduction). So we have concluded that the initial ECG represented an atrial tachycardia with left bundle branch block aberrancy conducted 1:1 and that the control ECG tracing which was thought to represent a sinus rhythm was atrial rhythm's with 2:1 atrioventricular conduction. The patient underwent cardiac magnetic resonance imaging (CMR). Cine images showed a huge right atrium, apical displacement of the tricuspid valve leaflets and normal systolic function of RV and LV **(Figure 6)**.

Radiofrequency ablation was not performed because it was judged not feasible. Combination therapy with Digitalis and Amiodarone therapy was instituted to control heart rate and for medical conversion. At two months follow up, the patient's cardiac condition worsened to the level of NYHA class IV despite a sinus rhythm and echocardiography revealed worsening tricuspid regurgitation with lack of leaflet coaptation. She underwent surgical repair with tricuspid valve and a combined total cavopulmonary connection with fenestrated RV exclusion technique. She was discharged in good hemodynamic condition with an ejection fraction of 57%, and

she remained well with an improved of NYHA class in sinus rhythm on amiodarone.

3. DISCUSSION

E.A is a rare congenital heart disorder occurring in ≈1 per 200.000 live births and accounting for <1% of all congenital heart diseases [1]; only 5% of patients survive beyond the fifth decade without surgical correction [2]. Adults usually present with progressive cyanosis, decreasing exercise tolerance, or arrhythmias [3-4]. In the present case, the 60-year-old patient presented with dyspnea NYHA functional class II and wide complex tachycardia. Ebstein anomaly is characterized by apical displacement of the septal or posterior leaflet of the tricuspid valve, resulting in 'atrialization' of the inflow tract of the right ventricle and consequently a variably small, but functional right ventricle. Varying degrees of tricuspid regurgitation result from this morphology with consequent right atrial enlargement. The diagnosis of this disease can often be made by echocardiography and apical displacement of the septal leaflet of the tricuspid valve by 8mm/m2 or more confirms the diagnosis [5]. In the present case, the apical displacement was approximately 10 mm/m2, which fulfilled the diagnostic criterion. Associated anomalies include atrial septal defect in approximately 80% to 94% of patients with E.A [6-7]. Additional associated anomalies include bicuspid or atretic aortic valves, pulmonary atresia or hypoplastic pulmonary artery, subaortic stenosis, coarctation, mitral valve prolapse, accessory mitral valve tissue or muscle bands of the left ventricle, ventricular septal defects, and pulmonary stenosis. [8]. But neither of these anomalies were observed in our patient.

Arrhythmias are common in patients with E.A. Furthermore, sudden death has been reported in 2.6 % of these patients [9] .Conventional surgical repair of the tricuspid valve could not prevent the occurrence of sudden death [10]. Paroxysmal tachyarrhythmias are based on typical, fast-conducting atrioventricular accessory pathways with both antegrade and retrograde conduction properties in most patients. [11]. In addition, wide QRS tachycardia over a septal accessory atrioventricular pathway, ventricular tachycardia (VT), or flutter, as well as ectopic atrial tachycardia, atrial flutter, and atrial fibrillation, can occur[11-12].

Pre-excitation is caused by the downward displacement of the septal leaflet of the tricuspid valve which is associated with discontinuity of the central fibrous body and septal atrioventricular ring with direct muscular connections, thus creating a potential substrate for accessory atrioventricular connections and pre-excitation. From 6% to 36% of patients with E.A have pre-excitation [11-12-13-14]. But neither of these accessory pathways was revealed in the present patient.

Atrial fibrillation and flutter are most likely caused by the tricuspid valve insufficiency which results in right atria1 dilatation and a predilection for premature atrial contractions, in other hand it can be caused by secondary alterations of the right atrial myocardium from previous cardiac surgery or are postoperative as a result of incisional atrial tachycardia. [11].

According to these results, most reports in the literature considered the resulting supraventricular tachyarrhythmia as a cause of palpitation and/or sudden death in this anomaly. The role of ventricular tachyarrhythmia in sudden death in patients with E.A has not been delineated. Although it has been reported that during cardiac catheterization manipulation of the catheters in the atrialized right ventricle might result in VT and ventricular fibrillation, Kastor et al [15] systematically studied the electrophysiologic characteristics of patients with E.A. The results showed that the atrialized RV was particularly irritable. The anatomic basis for the development of VT in E.A might be supported by histopathologic studies that show that the myocardium in the right ventricle of patients with E.A is morphologically abnormal [16]. But, in our case, the programmed ventricular stimulation was negative.

The management of tachyarrhythmias associated with E.A remains a real challenge [17]. Electrophysiological evaluation and radiofrequency ablation of symptomatic supraventricular tachyarrhythmia should be performed when feasible.

But it remains difficult because of atrial dilatation, which disrupts anatomic landmarks and makes it difficult to find and target the atrioventricular junction [18]. In our case, radiofrequency ablation of the multiple ectopic atrial foci was judged not feasible because of marked enlargement of the right atrium and the severe tricuspid regurgitation which result in instability of the ablation catheter. Supraventricular tachyarrhythmia associated with E.A also can be ablated at the time of operative repair [19-20]. But the results are less impressive in cases of atrial flutter or fibrillation.

CONCLUSION

There are many mechanisms of tacchyarythmia in E.A and their management remains a real challenge. In our case regular wide QRS complexes tachycardia was suggestive of ventricular tachycardia. ECG after interruption of tachycardia mimicked a sinus rhythm. The diagnosis of 1/1 atrial tachycardia was retrospectively made. The surgical correction of tricuspid regurgitation was associated with the clinical improvement of the patient and the arrhythmia was only treated with amiodarone.

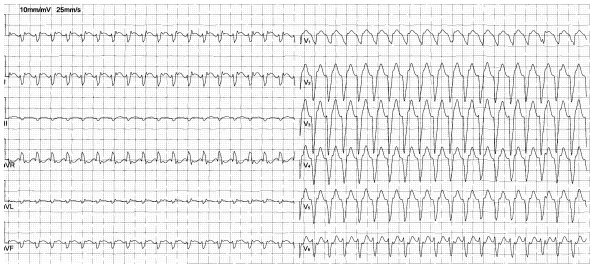

Figure 1: The 12-lead ECG during tachycardia: wide QRS tachycardia with left bundle branch (LBB) morphology and left axis deviation

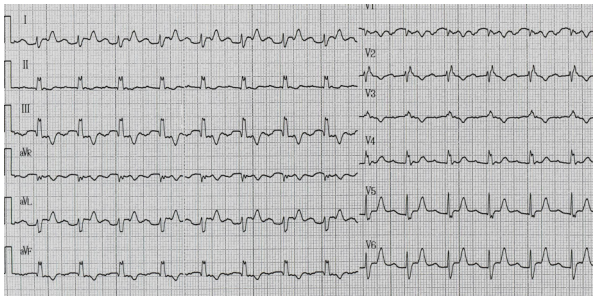

Figure 2: ECG after interruption of tachycardia

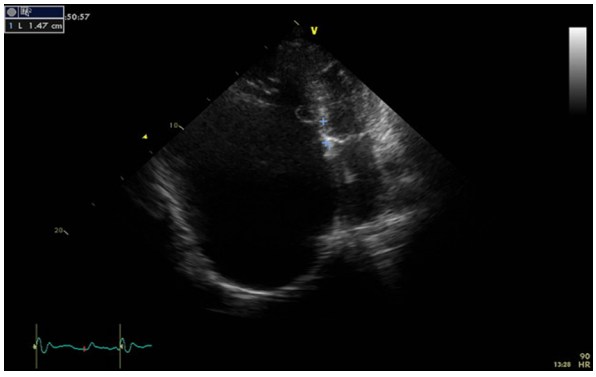

Figure 3: 4 Transthoracic echocardiography; apical 4 chambers view ; apical displacement of the septal and posterior leaflets of the valve (15mm/m2) and marked enlargement of the right atrium

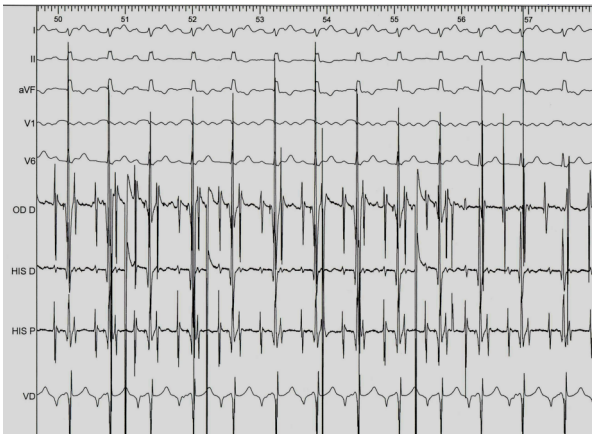

Figure 4: Intracardiac recordings indicated an atrial tachycardia with 2:1 conduction.

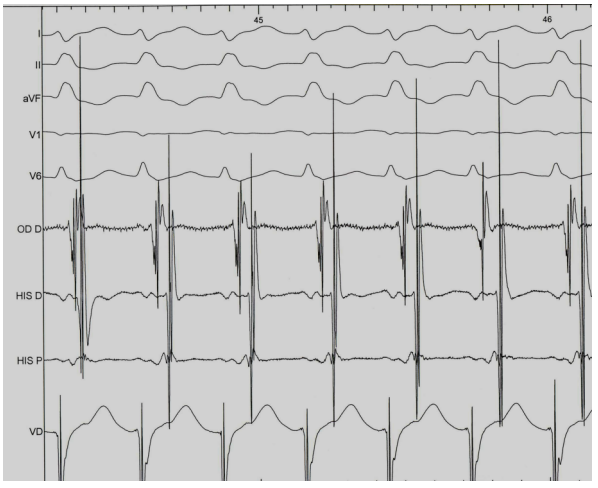

Figure 5: 1:1 atrial tachycardia during isoproterenol infusion

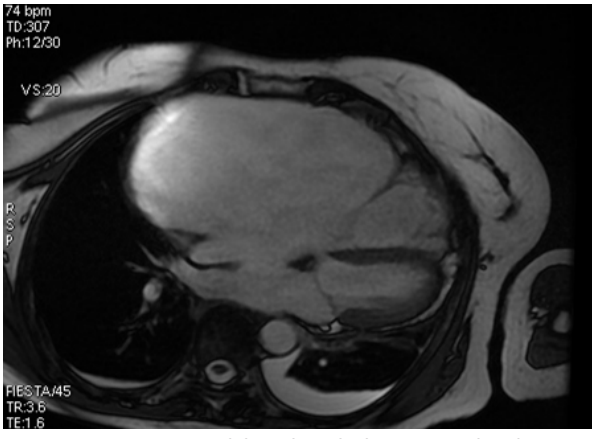

Figure 6: Cine CMR at end diastole in the long-axis 4-chamber View (3T scanner GE): Normal RV systolic function with large RA enlargement.

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