A Young Woman With Recurrent Palpitations: A Case of Ebstein Anomaly With Mahaim Fiber Tachycardia

Michael C. Viray, MD, Philip C. Wiener, DO, Uyanga Batnyam, MD, Lorena Rasquin, MD, Gregg S. Pressman, MD, FACC, and Sumeet Mainigi, MD, FHRS, *Philadelphia, Pennsylvania*

INTRODUCTION

We present the interesting case of a young woman presenting with recurrent palpitations who was found to have Ebstein anomaly (EA) on transthoracic echocardiography with associated Mahaim fiber tachycardia on comprehensive electrophysiologic study (EPS). We discuss the pivotal role of echocardiography in clinching the diagnosis of EA and its influence on guiding appropriate therapy.

CASE PRESENTATION

A 26-year-old woman with a medical history of hyperthyroidism presented to our institution with acute onset of shortness of breath and palpitations. She reported on-and-off palpitations for the past 8 years that contributed to anxiety attacks. She was hemodynamically stable on presentation, without signs of volume overload. Her initial 12-lead electrocardiogram demonstrated a wide-complex tachycardia at 178 beats/min with a left bundle branch block morphology (Figure 1), which raised suspicion for the presence of supraventricular tachycardia with aberrancy versus ventricular tachycardia. The arrhythmia was terminated by a single dose of adenosine.

Bedside transthoracic echocardiography revealed an apically displaced tricuspid valve with a redundant anterior leaflet, a dilated right atrium with a small functional right ventricle, mild tricuspid regurgitation, and severe diffuse left ventricular systolic dysfunction (Videos 1-4). In addition, she possessed a unique M-mode tracing consistent with EA (Figure 2).

Her severe left ventricular dysfunction was suspected to be related to tachycardia-mediated cardiomyopathy, and therefore she was started on metoprolol succinate and lisinopril. In addition, because of a strong suspicion for the presence of an accessory pathway, she subsequently underwent a comprehensive EPS, which revealed findings consistent with an atrioventricular reentrant tachycardia with antidromic conduction (Figure 3). Intracardiac electrogram demonstrated identical pattern showing "His-like" signal consistent with Mahaim and corroborated with catheter placement on fluoroscopy (Figure 4). Three-dimensional electrocardiographic activation mapping of the right atrium and right ventricle elucidated the diagnosis of antidromic Mahaim tachycardia, which is antegrade conduction using an atriofascicular pathway along the lateral wall and retrograde conduction over the atrioventricular (AV) node (Figure 5). Subsequent radiofrequency ablation (RFA) of the accessory pathway resulted in a right bundle branch block pattern typical of EA on electrocardiography (Figure 6). The patient was discharged home with a LifeVest (Zoll Medical, Chelmsford, MA) and referred to an adult congenital heart disease center for further management.

DISCUSSION

EA is a rare congenital heart disorder that accounts for <1% of all cases of congenital heart disease (about one per 200,000 live births) and was first described by Wilhelm Ebstein in 1886 at autopsy of a young man who had presented with recurrent palpitations and dyspnea since childhood.[1,2] It is characterized by apical displacement of the septal and often posterior leaflet of the tricuspid valve into the right ventricular cavity. Embryologically, it arises from failure of the septal and posterior tricuspid valve leaflets to delaminate from the underlying myocardium at the inlet portion of the right ventricle to which they are adherent. Consequently, the right ventricle is divided into a proximal "atrialized" portion and a distal "functional" portion.[3] EA is usually associated with other congenital cardiac anomalies, such as atrial septal defects (most commonly), ventricular septal defects, pulmonic stenosis, tetralogy of Fallot, coarctation of the aorta, and mitral valve abnormalities.[4]

Adult patients with undiagnosed EA usually present with arrhythmias and symptoms of congestive heart failure such as fatigue, poor exercise tolerance, and exertional dyspnea. Up to 30% of patients with EA have accessory pathways, and most of these are located around the orifice of the malformed tricuspid valve.[5] It is thought that structural and histologic abnormalities around the right atrioventricular junction and anatomic tricuspid valve form the substrates for these reentrant arrhythmias.[6] Ventricular arrhythmias are rare in EA despite the significant ventricular dilatation and underlying abnormal myocardium.

Echocardiography is the key diagnostic modality in EA, providing information on the anatomy and function of the tricuspid valve, the right ventricular outflow tract, the associated congenital cardiac lesions, and the size and function of the ventricles. It also provides useful data to assess suitability for tricuspid valve repair or replacement in patients with EA. Echocardiographic indicators of poor surgical outcomes include increased preoperative right ventricular size, severe tethering of the anterior mitral leaflet, severely deformed septal/posterior leaflet, presence of right ventricular outflow tract obstruction, presence of preoperative left ventricular dysfunction, and presence of preoperative right ventricular dysfunction.[7]

Success rates of RFA of accessory pathways are typically lower in patients with EA than in patients with structurally normal hearts. It

is important to perform selective right coronary angiography to help identify the true tricuspid annulus, which is the ideal site of ablation. Timing of invasive EPS and RFA is ideally done preoperatively, as surgical therapies such as tricuspid valve replacement and cone reconstruction of the tricuspid valve limit postoperative access to ideal sites for transcatheter ablation.[8]

This patient underwent a comprehensive EPS, which revealed an interesting finding suggestive of an AV reentrant tachycardia with antidromic conduction via a Mahaim pathway, which is antegrade conduction using an atriofascicular pathway and retrograde conduction over the AV node. Mahaim pathways are an extremely rare cause of reentrant arrhythmias. They constitute <3% of accessory pathways but are frequently associated with EA. Mahaim fibers have AV node–like conduction properties whose proximal insertions are localized in the lateral, anterolateral, or posterolateral part of the tricuspid annulus and whose distal fibers connect to the right ventricular free wall or the right bundle branch. Adenosine sensitivity and decremental conduction along these anomalous connections is what distinguishes these fibers from the majority accessory pathways. Subsequent RFA of the
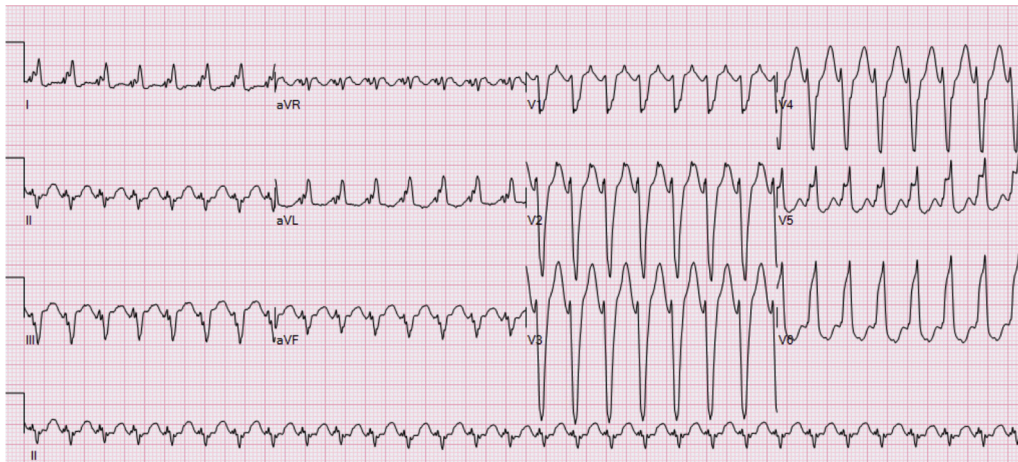

Figure 1 Initial presenting 12-lead electrocardiogram demonstrating a wide-complex tachycardia at 178 beats/min with a left bundle branch block morphology.
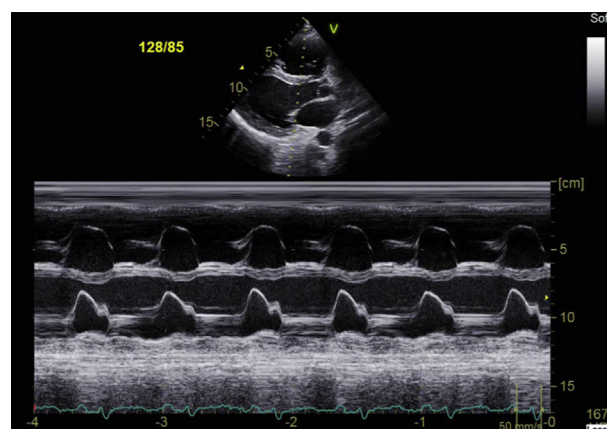

Figure 2 A characteristic M-mode tracing for Ebstein Anomaly demonstrating delayed closure of the tricuspid valve compared with the mitral valve.
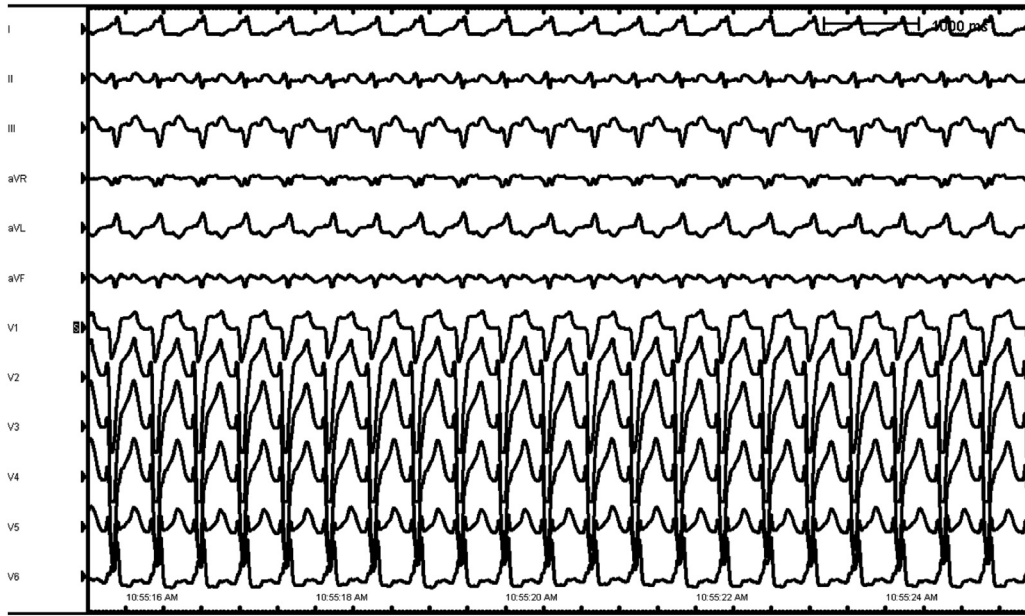
Figure 3 Induced arrthymia during electrophysiology study reproduces clinical tachycardia.
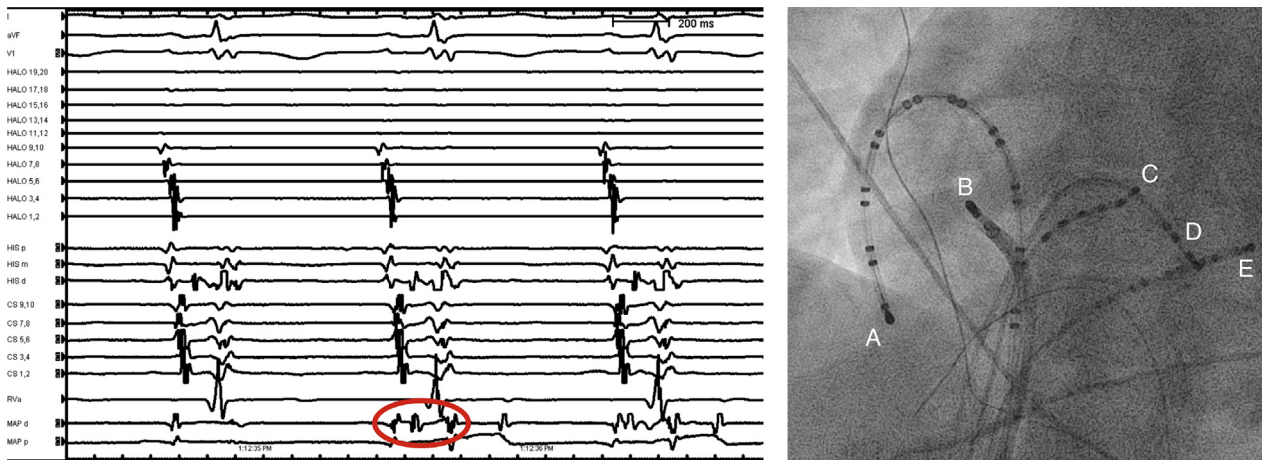

Figure 4 Intracardiac electrogram demonstrating identical "His-like" pattern in mapping catheter (*red circle*) consistent with Mahaim tachycardia (*left*) and confirmed with concomitant RF catheter placement on the lateral wall under fluoroscopy (*right*). (**A**) "Halo" duo-decapolar catheter; (**B**) radiofrequency catheter; (**C**) his catheter; (**D**) RV apex catheter; (**E**) coronary sinus catheter.
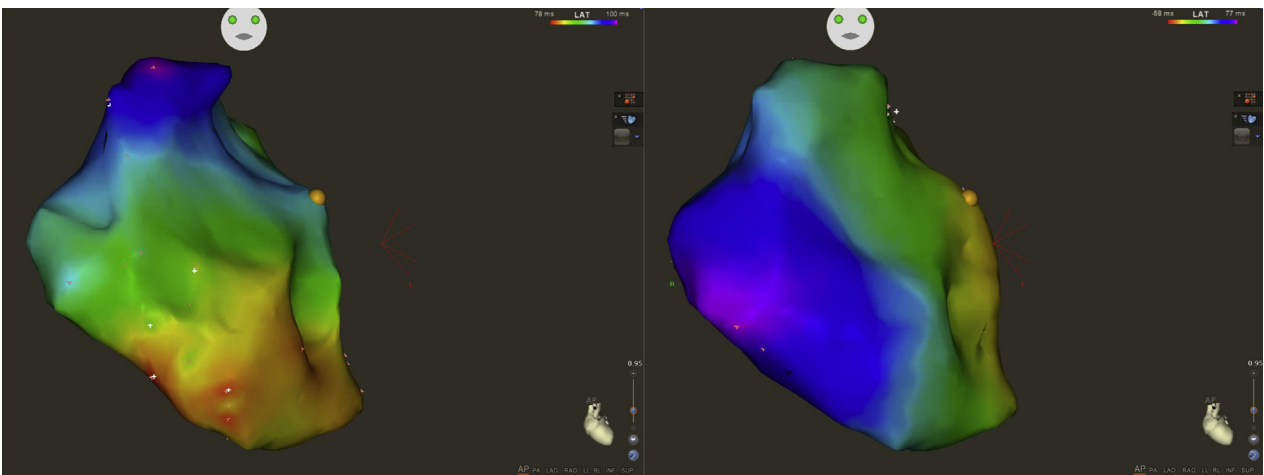

Figure 5 Three-dimensional electrocardiographic activation mapping visualizing antegrade conduction using an atriofascicular pathway on the lateral wall (*left*) and retrograde conduction through the AV node on the septal wall (*right*).
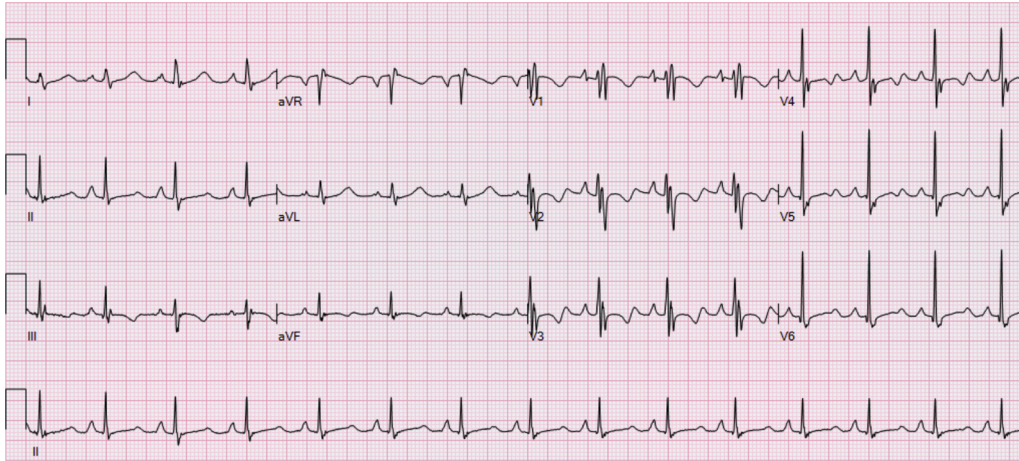
Figure 6 Twelve-lead electrocardiogram obtained following successful RFA of the accessory pathway demonstrating a right bundle branch block pattern of typical of EA.

Mahaim pathway resulted in development of a right bundle branch block pattern on intracardiac electrocardiography, another finding that is characteristic of Mahaim fiber tachycardias.

CONCLUSIONS

Echocardiography played a pivotal role in revealing an uncommon congenital cardiac abnormality. Through a multimodal approach, we described the case of a young adult woman with recurrent palpitations noted to have EA, an uncommon congenital cardiac abnormality, revealed by echocardiography. In addition, our patient demonstrated on EPS a Mahaim fiber tachycardia, which is rare in the general population but more prevalent in patients with EA.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.03.006.

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