

Wide complex tachycardia in an elderly woman due to Ebstein's anomaly with two accessory pathways



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Introduction

Among elderly patients presenting with wide complex tachycardia, the differential diagnosis is skewed toward ventricular tachycardia (VT), given their higher prevalence of structural heart disease. Despite this, other diagnoses should be considered, including late presentations of congenital anomalies. The aim of this case report is to describe a de novo diagnosis of Ebstein's anomaly (EA) in an elderly patient presenting with wide complex tachycardia. Her initial electrocardiogram (ECG) was consistent with pre-excited atrial fibrillation (AF), and once in sinus evidence of multiple accessory pathways was present. Imaging confirmed the diagnosis of EA, and she ultimately underwent successful catheter ablation.

Case report

A 76-year-old woman with a history of hypertension, hyperlipidemia, subdural hemorrhage, and coronary artery disease presented to the emergency room with chest pressure, nausea, and diaphoresis. In the field, she was found to be in a wide complex tachycardia at a rate of 220 beats per minute (bpm) with a systolic blood pressure of 70 mm Hg. Her initial ECG is shown (Figure 1A). In the emergency room, she was diagnosed with unstable VT, loaded with intravenous amiodarone 150 mg bolus twice, and cardioverted at 200 J. She was then intubated. Subsequent ECG in the emergency room showed sinus rhythm with a short PR interval and a delta wave (Figure 1B). The patient was started on an amiodarone infusion and transferred to the cardiac care unit for further management. It was suspected that the initial irregular wide complex tachycardia was pre-excited AF. However, her history of coronary disease, age, and predominance of precordial negativity during tachycardia suggested a possibility of VT.

KEYWORDS Accessory pathway; Ebstein's anomaly; Elderly; Irregular tachycardia; Pre-excitation; Wide complex tachycardia (Heart Rhythm Case Reports 2019;5:205–208)

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KEY TEACHING POINTS

- Despite risk factors, not all wide complex tachycardias are ventricular tachycardia, and judicious attention to electrocardiograms and imaging may reveal other pathologies.
- Diagnosis of Ebstein's anomaly is made via imaging of apical displacement of the septal tricuspid leaflet ≥ 0.8 cm/m² relative to the anterior mitral leaflet insertion.
- Nearly 50% of patients with Ebstein's anomaly have multiple accessory pathways.

She underwent invasive cardiac angiography, which found a discrete 40% stenosis of the proximal right coronary artery and a left ventricular end diastolic pressure of 21 mm Hg. Her transthoracic echocardiogram (TTE) showed a left ventricular ejection fraction of 45% (Figure 2A and B). Cardiac magnetic resonance imaging also showed mild left ventricular systolic dysfunction, an enlarged right atrium, and an apically displaced tricuspid valve (TV) without late gadolinium enhancement. Subsequent cardiac computed tomography confirmed EA with a posterior TV leaflet displaced 1.7 cm/m² (26 mm) toward the right ventricular apex from the insertion point of the anterior mitral valve leaflet, resulting in an atrialized region of the right ventricle (Figure 2C–F).

On electrophysiologic (EP) testing, she was found to have an HV interval of 15 ms. VT could not be induced with triple extrastimuli from 2 different sites. No supraventricular tachycardia was induced, but rapid atrial pacing induced AF with the morphology of the presenting ECG. This confirmed the diagnosis of pre-excited AF. Electroanatomic mapping during atrial pacing mapped the earliest ventricular activation to the posterolateral tricuspid annulus (Figure 3A and B). Ablation at this site led to a change in the QRS morphology (Figure 3C). There was now an isoelectric delta in V₁ and a

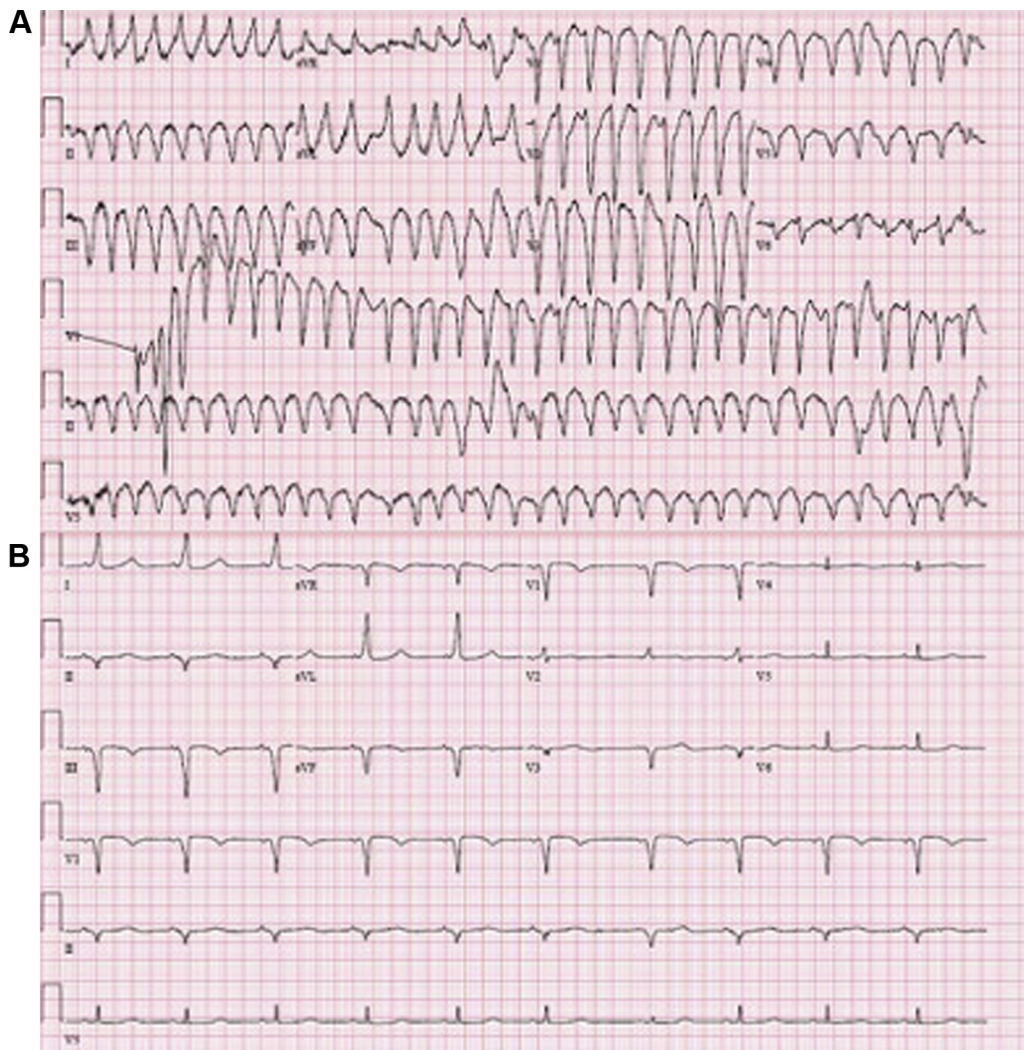


Figure 1 A: Presenting electrocardiogram (ECG). B: ECG post cardioversion showing delta wave and short PR interval.

positive QRS, indicative of a septal accessory pathway. The septal tricuspid annulus was mapped, and pre-delta activation was noted; however, prior to ablation it was decided to map the left side to assess for earlier activity. A single transeptal puncture was performed under intracardiac ultrasound guidance. The posterior and septal mitral annulus were mapped but did not have pre-delta activation. Ablation was then performed at the posteroseptal tricuspid annulus, which led to termination of the second pathway (Figure 3C). Adenosine testing revealed no further evidence of an accessory pathway. The ECG post ablation of both pathways is shown in Figure 3D. An implantable loop recorder was placed at discharge; there has been no AF at up to 1 year of follow up.

Discussion

Our case is unique owing to the advanced age at presentation, history of coronary artery disease, irregular monomorphic tachycardia with mostly fixed pre-excitation, and different pre-excitation pattern in sinus rhythm and AF. While the

irregularity of the wide complex tachycardia and the delta wave on the sinus rhythm ECG strongly suggested the initial presentation was pre-excited AF, the history of coronary disease, advanced age, mildly reduced ejection fraction, and predominance of precordial negativity raised the specter of VT. During the EP study, supraventricular tachycardia or VT was not inducible, but AF was induced and mimicked the activation pattern of the initial ECG. Comparison of the ECG in AF and sinus showed evidence of dual accessory pathways. The initial ECG in AF (Figure 1A) shows a late precordial transition suggestive of a free wall pathway, while the ECG in sinus rhythm shown in Figure 1B shows a positive delta wave in lead I and an isoelectric delta wave in V_1 with $R < S$, suggesting a septal accessory pathway.

The ECG in AF, which showed a relatively fixed pattern of preexcitation, suggests preferential conduction via the posterolateral pathway. During electrophysiologic testing, we found that the septal pathway had a longer refractory period (320 ms) than the posterolateral pathway (210 ms). Thus, during AF there was preferential conduction down

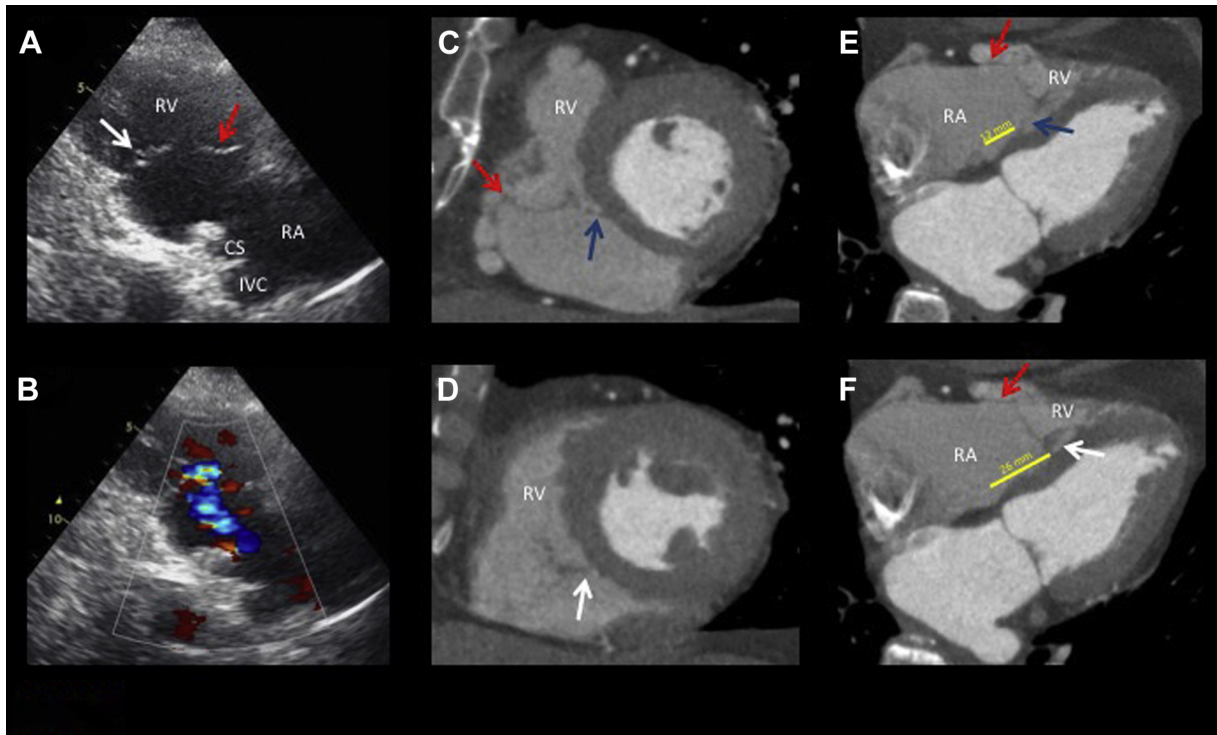


Figure 2 **A:** Transthoracic echocardiogram right ventricular inflow view demonstrates apical displacement of the posterior tricuspid valve leaflet (*white arrow*). The *red arrow* marks the anterior tricuspid valve leaflet. The area between the visualized tricuspid valve leaflets and the coronary sinus (CS) represents atrialized right ventricle (RV). **B:** Color flow Doppler in the same plane shows a jet of tricuspid regurgitation. **C, D:** Cardiac computed tomography in the short-axis view at the basal (C) and mid (D) ventricular levels shows the anterior (*red arrow*), septal (*blue arrow*), and posterior (*white arrow*) tricuspid valve leaflets. **E, F:** The 4-chamber view demonstrates a 12-mm and 26-mm displacement of the septal (E) and posterior (F) tricuspid valve leaflets, respectively, from the anterior mitral valve leaflet insertion. *Red, blue, and white arrows* mark the same tricuspid valve leaflet (anterior, septal, or posterior) throughout the figure. IVC = inferior vena cava; RA = right atrium; RV = right ventricle.

the posterolateral accessory pathway with concealed retrograde penetrance in the posteroseptal pathway making the posteroseptal pathway refractory to anterograde conduction.

With regard to the posteroseptal pathway, intracardiac mapping of the septum on the tricuspid annulus initially led to pre-delta activation (-20 ms pre-delta). It is rare that patients with EA have left-sided pathways, but given the isoelectric delta, and $R < S$ in V_1 , which can be consistent with a septal tricuspid or mitral pathway, it was decided to map the mitral annulus prior to ablation.¹ Left-sided activation was on time with the delta wave, and thus further mapping along the tricuspid annulus was performed. Successful termination was achieved on the posteroseptal tricuspid annulus (-25 ms pre-delta).

EA is a rare congenital heart defect (5 in 100,000 live births) resulting in a spectrum of TV and right ventricular abnormalities.^{2–6} Abnormalities in EA range from minimal displacement of the septal and posterior leaflets to significant displacement with tethering and severe regurgitation. In turn, clinical presentations can vary from hydrops fetalis in the neonate to incidental detection in adulthood. Delamination of tricuspid valve leaflets leads to apical displacement, dilatation of the atrialized portion of

the right ventricle, dilatation of the anatomic TV annulus, and tricuspid regurgitation.²

In adults, EA typically presents with arrhythmias.⁵ Previous cases of EA presenting with wide complex tachycardia found to be pre-excited AF have been reported in middle-aged patients.⁷ Arrhythmias consist of accessory pathways (15%–20%) and AF or atrial flutter—the frequency of which increases with age (30%–40% in patients over the age of 50).^{2–4,8,9} Accessory pathways are attributed to the embryologic malformation and are therefore predominantly right-sided. These are most commonly posteroseptal by the coronary ostium and posterolateral concordant with the posterior and septal leaflets.¹⁰ It is important to note that accessory pathways occur at the morphologic atrioventricular annulus, not at the functional annulus. Almost 50% of patients with EA have multiple accessory pathways. Of note, VT is rare in these patients.⁵

Diagnosis is made by noninvasive imaging. TTE is the initial test of choice. Characteristic signs are apical displacement of the septal tricuspid leaflet ≥ 0.8 cm/m² relative to the anterior mitral leaflet insertion. The posterior leaflet can also be displaced in more severe cases. The most consistent finding is an enlarged right atrium. Magnetic resonance imaging is the gold standard.^{2,3}

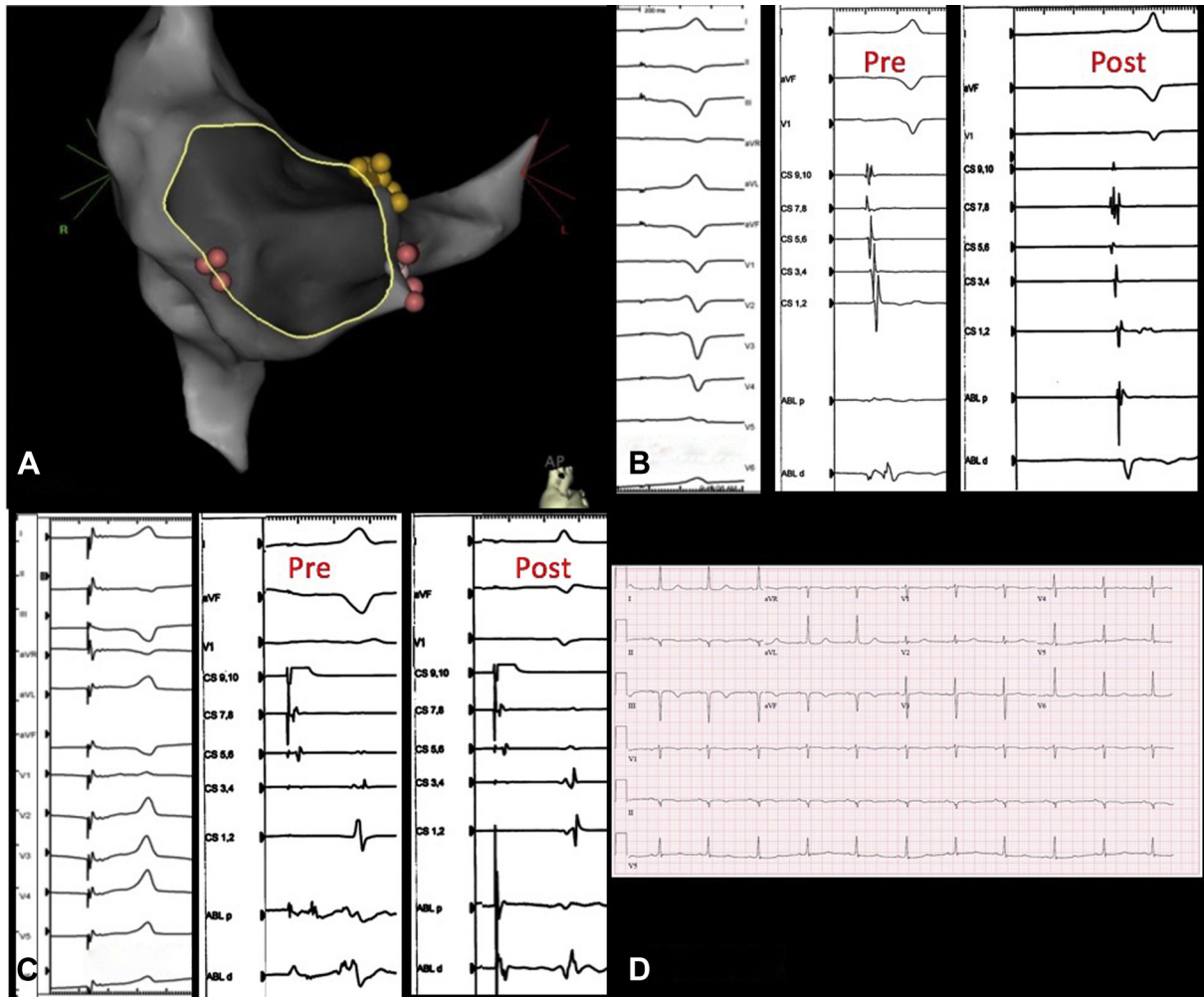


Figure 3 A: Electroanatomic mapping in left anterior oblique view of right atrium. Yellow tags indicate location of His potentials. Two sites of accessory pathways are noted by red tags. B: Surface electrocardiogram (ECG) and local electrograms at site of posterolateral pathway before (Pre) and after (Post) ablation. C: Surface ECG and local electrograms at site of posteroseptal pathway before (Pre) and after (Post) ablation. D: Surface ECG at end of case. (Speed: 25 mm/s.) Note loss of pre-excitation. AP = anteroposterior; L = left; R = right.

Conclusion

We present a unique case of EA in an elderly patient. While the irregular wide complex tachycardia and delta wave suggested pre-excited AF, the patient's advanced age, history of coronary disease, and precordial negativity raised the possibility of VT. A combination of advanced cardiac imaging and EP study was used to establish the diagnosis. Differences between the pattern of pre-excitation in AF and sinus rhythm ECG suggested 2 accessory pathways, which was confirmed during electroanatomic mapping.

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