Preexcitation syndrome unveiling Ebstein's anomaly at an exceptionally advanced age: A case report

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Case Report

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Abstract

**Background:** Ebstein's anomaly represents an infrequent congenital cardiac anomaly characterized by atypical tricuspid valve morphology, accompanied by the displacement of the septal and posterior leaflets towards the right ventricle. This condition is commonly associated with a variety of arrhythmias, including atrial fibrillation. Pre-excited atrial fibrillation denotes a distinct subtype of atrial fibrillation in which an accessory pathway bypasses the standard atrioventricular conduction system, consequently leading to rapid ventricular response rates.

**Case presentation:** We present a unique instance of atrial fibrillation with preexcitation revealing Ebstein's anomaly in an elderly patient. The patient, a 71-year-old individual with no previous medical or surgical history, presented with symptoms of palpitations and shortness of breath. The clinical examination revealed a rapid tachycardia that was poorly tolerated hemodynamically. Electrocardiogram findings demonstrated characteristic features of preexcited atrial fibrillation, which were subsequently confirmed by an electrophysiological study, indicating the presence of an accessory pathway. Echocardiography revealed the typical manifestations of Ebstein's anomaly. Initially, the patient underwent urgent cardioversion, which successfully restored sinus rhythm. Since the ablation procedure was unsuccessful, the patient's treatment approach was modified to focus on medical management, which ultimately resulted in a favorable outcome.

**Conclusions:** This unique case sheds light on the complex interplay between cardiac arrhythmias and adult congenital heart disease. Early diagnosis and comprehensive management are crucial in optimizing outcomes and improving the quality of life for patients with this rare combination of cardiac abnormalities. Further research is warranted to better understand the optimal treatment strategies, and long-term outcomes in this specific population.

Introduction

Ebstein’s anomaly, initially described by Wilhelm Ebstein in 1866, is a rare congenital malformation characterized by significant abnormalities primarily affecting the tricuspid valve and the right ventricle (1). Depending on the extent of anatomical abnormalities, the clinical presentation of Ebstein anomaly can vary widely, ranging from critically ill fetuses to asymptomatic adults. It affects approximately 1 in 20,000 live births in the general population and represents less than 0.5% of all reported cases of congenital heart disease (2). Cases of syncope and sudden death have been documented and can potentially be attributed to atrial fibrillation with a rapid ventricular response resulting from accelerated conduction via an accessory pathway, or to ventricular arrhythmias. Indeed, Accessory pathways are found in approximately 15–25% of cases, with more than 90% of these pathways being located on the right side. Among these cases, about 30% involve multiple accessory pathways, and around 10% exhibit atypical conduction properties (3). Numerous publications have documented cases of Ebstein's anomaly associated to atrial arrhythmias, especially in young people, as severe cases can lead to early mortality. To the best of our knowledge, it's the first case of pre-excited atrial fibrillation as the revealing mode of Ebstein's anomaly in a 71-year-old individual.

Our paper was written according to the CARE guidelines. (4)
Case presentation

A 71-year-old female patient with no previous medical or surgical history presented to the emergency department with complaints of palpitations and shortness of breath. The patient reported intermittent episodes of palpitations over the past few years but had never sought medical attention for them. She presented with hemodynamic instability characterized by hypotension measuring 80/30mmHg and a heart rate approximately at 175 beats per minute (bpm). Physical examination revealed an irregularly irregular pulse, without any signs of heart failure. An electrocardiogram (ECG) was performed, which demonstrated a wide-complex tachycardia that was irregularly irregular, displaying varying morphologies from one complex to another without significant alterations in the electrical axis. This pattern created an accordion-like appearance, which strongly indicated pre-excited AF (Figure 1). Thus, an urgent electrical cardioversion was performed, leading to the successful restoration of sinus rhythm. The ECG in sinus rhythm revealed a shortened PR interval and a negative delta wave in V1, along with a QRS transition occurring before V3. Additionally, the most prominent positive delta wave was observed in AVL, which suggests the presence of a right posteroseptal Kent bundle (Figure 2). Transthoracic echocardiography was subsequently performed to evaluate the underlying etiology of the patient's arrhythmia. The results revealed a structurally abnormal tricuspid valve, characterized by enlarged and redundant leaflets and apical displacement of the septal and posterior leaflets (Figure 3-A and 3-B). Furthermore, a portion of the right atrium was incorporated into the right ventricle, resulting in an "atrialization" of the right ventricle and severe right atrial enlargement (Figure 3-C). The right ventricle (RV) appeared small and did not show decreased systolic function (Figure 3-E and 3-F). Additionally, severe tricuspid regurgitation was observed (Figure 3-D). To investigate for other associated cardiac abnormalities such as atrial septal defect (ASD) and patent foramen ovale (PFO), transesophageal echocardiography was performed, but no remarkable findings were detected. An electrophysiology study (EPS) was conducted to identify the exact location and characteristics of an accessory pathway (Figure 4). Unfortunately, the localization of the accessory pathway (AP) was impossible due to a massively enlarged right heart, displaced tricuspid annulus, and distortion of anatomical landmarks, all of which made catheter stability difficult.

After a multidisciplinary consultation within a Heart team, taking into account the patient's age and her expressed preference, it was decided to initiate a medical treatment involving the administration of flecainide, beginning with a dosage of 50mg twice daily. This approach deliberately excludes other antiarrhythmic agents known to induce atrioventricular (AV) node blockade, such as beta-blockers, verapamil and amiodarone. Additionally, the patient was prescribed anticoagulation therapy with Rivaroxaban 20mg per day as a measure for stroke prevention. Diuretics were also prescribed to manage tricuspid regurgitation. Regular follow-up visits were scheduled to monitor the patient's clinical status and evaluate the necessity for any additional interventions. During these visits, no notable abnormalities or concerns were identified.

Discussion

Ebstein's anomaly is a congenital heart defect that typically manifests in early childhood, making its discovery in adulthood a rare occurrence. In a retrospective cohort study conducted at Mayo Clinic between 2003 and 2020, examining adults with Ebstein's anomaly, the age range of the 682 patients included in the
analysis was between 24 and 49 years (5). However, our presented case is distinct, as the patient was first diagnosed with Ebstein's anomaly at the age of 71 years old, and the presence of preexcitation served as the revealing mode for this condition. This highlights the rarity and atypical nature of our case, considering the advanced age at diagnosis and the unique clinical presentation. The diagnosis of Ebstein's anomaly is established through the identification of apical displacement of the attachment point of the septal tricuspid valve leaflet, relative to the attachment point of the anterior mitral valve leaflet, as demonstrated in the apical four-chamber transthoracic echocardiographic view. This displacement is measured and indexed by body surface area, with a threshold of $\geq 8$ mm/m$^2$ (6). The identification of Ebstein's anomaly in this case was unexpected given the patient's age and background. Cardiac magnetic resonance imaging (CMR) provides valuable information that complements echocardiography findings, offering additional insights into the anatomy of the tricuspid valve as well as the quantification of right ventricular size and function. In our specific case, considering the potential for severe tricuspid regurgitation to overestimate right ventricular function, CMR would have constituted an outstanding choice. The underlying mechanisms for the development of atrial fibrillation in Ebstein's anomaly are not well understood but are thought to be related to atrial enlargement, abnormal electrical conduction pathways, and structural changes within the atria (5). Apart from the hemodynamic strain imposed by the underlying valve abnormality, individuals with Ebstein's anomaly also face an exceptionally elevated prevalence of tachyarrhythmias, primarily caused by the presence of accessory atrioventricular pathways (APs) situated adjacent to the posterior and septal margins of the tricuspid valve, where the valve leaflets exhibit the most pronounced abnormalities (7). Our case is consistent with these findings, as the implementation of the EASY-WPW algorithm (8) enabled us to accurately locate the accessory pathway in the right posteroseptal region. This finding was subsequently confirmed through an electrophysiology study. The late-onset diagnosis of Ebstein's anomaly in this patient brings forth significant implications. Firstly, it underscores the necessity for healthcare professionals to maintain a heightened level of suspicion for uncommon cardiac conditions, even in older individuals without a congenital heart disease background. Secondly, it highlights the significance of thorough cardiac assessment, encompassing echocardiography, in patients presenting with atrial fibrillation or other cardiac arrhythmias. This is especially crucial when these individuals display atypical characteristics or exhibit a lack of response to conventional treatment strategies. Furthermore, the clinical management of atrial fibrillation with preexcitation in the setting of Ebstein's anomaly presents several challenges. The primary concern is the choice of antiarrhythmic therapy. In this context, the use of AV node blockers, including medications like adenosine, beta-blockers, verapamil, and amiodarone, needs to be carefully considered. This is because these medications can block the normal electrical pathway through the AV node, thereby forcing the electrical impulses to use the accessory pathway increasing the risk of rapid ventricular response, which can lead to hemodynamic instability and adverse outcomes (8, 9). The complex anatomical features associated with Ebstein's anomaly, such as a significantly enlarged right heart, a displaced tricuspid annulus, and distorted anatomical landmarks, can pose increased difficulties during catheter ablation procedures (10). Achieving and maintaining catheter stability can be challenging due to these factors, as we experienced in our case. The surgical intervention plays a pivotal role in the management of Ebstein's anomaly among adult patients. Tricuspid valve repair is frequently favored, aiming to restore the valve's normal function and minimize the requirement for artificial valves (6). Heart transplantation may be considered as a final option in rare instances where there is severe right ventricular dysfunction (6). Customized treatment strategies,
interdisciplinary care, and regular long-term monitoring are crucial in optimizing outcomes for adult patients with Ebstein's anomaly who undergo surgical treatment. In our case, the main issue pertaining to Ebstein's anomaly was related to rhythmic abnormalities. Therefore, following a multidisciplinary discussion within the Heart team and considering the patient's preference, it was decided to put the patient on medical treatment.

**Conclusion**

This case report highlights the importance of considering rare cardiac conditions, such as Ebstein's anomaly, even in individuals of advanced age. The coexistence of atrial fibrillation with preexcitation in this case adds further complexity to the clinical presentation. A multidisciplinary approach involving cardiologists, electrophysiologists, and imaging specialists is essential for accurate diagnosis and optimal management of these rare and challenging cases.

**Declarations**

**Availability of Data and Materials**

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

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The authors declare that they have no competing interests

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Figures
Figure 1

ECG findings: Accordion-like morphology with an irregularly irregular wide-complex tachycardia suggestive of pre-excited atrial fibrillation
Figure 2

The ECG after electrical cardioversion revealing a shortened PR interval and a negative delta wave in V1, along with a QRS transition occurring before V3 and with the most prominent positive delta wave was observed in AVL, which suggests the presence of a right posteroseptal Kent bundle.
Figure 3

Transthoracic Echocardiogram Results: (A-B): Apical four-chamber view reveals significant apical displacement of the septal tricuspid leaflet (orange arrow) relative to the anterior mitral leaflet (green arrow), leading to atrialization of the right ventricle. (C): Apical four-chamber view demonstrates pronounced enlargement of the right atrium. (D): Color-flow Doppler shows evidence of severe tricuspid valve regurgitation. (E-F): Longitudinal systolic function of the right ventricle appears normal based on tricuspid annular plane systolic excursion, S’ wave and isovolumic acceleration.
Figure 4

Position of catheters during an EPS. We note the massively enlarged right heart with the displaced tricuspid annulus and the distortion of anatomical landmarks, making the procedure difficult.