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Case Report

Pre-excitation Syndrome Unveiling Ebstein Anomaly at an Exceptionally Advanced Age

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Ebstein anomaly is a congenital cardiac anomaly that occurs infrequently. This condition is commonly associated with a variety of arrhythmias, including atrial fibrillation. We present a unique instance of atrial fibrillation with pre-excitation revealing Ebstein anomaly in an elderly patient. 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.

Ebstein 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 (RV).[1] Depending on the extent of anatomic abnormalities, the clinical presentation of Ebstein anomaly can vary widely, ranging from critically ill fetuses to asymptomatic adults. The anomaly 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 potentially can 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% to 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 anomaly that are associated with atrial arrhythmias, especially in young people, as severe cases can lead to early mortality. To the best of our knowledge, this case is the first of pre-excited atrial fibrillation as the revealing mode of Ebstein anomaly in a 71-year-old individual.

Our paper was written according to the CARE (**Ca**se **Re**ports) 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 having 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 80/30 mm Hg, and a heart rate of approximately 175 beats per minute. Physical examination revealed an irregularly irregular pulse, without any signs of heart failure. An electrocardiogram 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 atrial fibrillation (Fig. 1A). Thus, an urgent electrical cardioversion was performed, leading to the successful restoration of sinus rhythm. The electrocardiogram 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 lead AVL (augmented vector left), which suggests the presence of a right posteroseptal atriofascicular tract (Fig. 1B). Transthoracic echocardiography was performed subsequently to evaluate the underlying etiology of the patient's arrhythmia. The results

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Novel Teaching Points

- The late-onset diagnosis of Ebstein anomaly poses challenges, due to its rarity in older age groups.
- Difficulties in electrophysiological studies and catheter ablation procedures in the presence of complex anatomic features are associated with Ebstein anomaly.
- Ebstein anomaly poses management challenges, particularly the careful consideration of antiarrhythmic therapy in atrial fibrillation with pre-excitation, avoiding medications that may increase the risk of rapid ventricular response.
- This cases demonstrates the significance of a multidisciplinary approach involving cardiologists, electrophysiologists, and imaging specialists, for accurate diagnosis and optimal management of rare and challenging cases such as Ebstein anomaly in adults.

revealed a structurally abnormal tricuspid valve, characterized by enlarged and redundant leaflets and apical displacement of the septal and inferior leaflets (Fig. 2, A and B; Video 1, view video online). Furthermore, a portion of the right atrium was incorporated into the RV, resulting in an "atrialization" of the RV, and severe right atrial enlargement (Fig. 2C). The RV appeared small and did not show decreased systolic function (Fig. 2E). Additionally, severe tricuspid regurgitation was observed (Fig. 2D; Video 2, view video online). 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 (Fig. 2F). Unfortunately, localization of the accessory pathway was impossible, due to a massively enlarged right heart, a displaced tricuspid annulus, and distortion of anatomic landmarks, all of which made catheter stabilization difficult.

After a multidisciplinary consultation within a heart team, taking into account the patient's age and her expressed preference, the decision was made to initiate medical treatment involving the administration of flecainide, beginning with a dosage of 50 mg 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 20 mg per day as a measure for stroke prevention. Diuretics also were 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 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 of adults with Ebstein anomaly, conducted at the Mayo Clinic between 2003 and 2020, 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 anomaly at the age of 71 years, and the presence of pre-excitation served as the revealing mode for this condition. The rarity and atypical nature of our case are highlighted by the advanced age at diagnosis and the unique clinical presentation. The diagnosis of Ebstein anomaly is established through the identification of apical displacement of the attachment point of the septal and inferior tricuspid valve leaflets, relative to the attachment point of the anterior mitral valve leaflet, as demonstrated in the apical 4-chamber transthoracic echocardiographic view. This displacement is measured and indexed by body surface area, with a threshold of ≥ 8 mm/m^2.[6] The identification of Ebstein anomaly in this case was unexpected, given the patient's age and background. Cardiac magnetic resonance imaging provides valuable information that complements echocardiography findings, offering additional insights into the anatomy of the tricuspid valve as well as the quantification of RV size and function. In our specific case, considering the potential for severe tricuspid regurgitation to overestimate RV function, cardiac magnetic resonance imaging would have constituted an outstanding choice. The underlying mechanisms for the development of atrial fibrillation in Ebstein anomaly are not well understood, but they 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 anomaly also face an exceptionally elevated prevalence of tachyarrhythmias, caused primarily by the presence of accessory AV pathways situated adjacent to the septal and inferior margins of the tricuspid valve, where the valve leaflets exhibit the most pronounced abnormalities.[5] Our case is consistent with these findings, as the implementation of the EASY-WPW algorithm[7] 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 anomaly in this patient brings forth significant implications. First, 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. Second, it highlights the significance of thorough cardiac assessment, encompassing echocardiography, in patients presenting with atrial fibrillation or other cardiac arrhythmias. This assessment 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 pre-excitation in the setting of Ebstein anomaly presents several challenges. The primary concern is the choice of antiarrhythmic therapy. In this context, the use of AV node blockers, including medications such as adenosine, beta-blockers, verapamil, and amiodarone, needs to be considered carefully. The reason for this is that 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

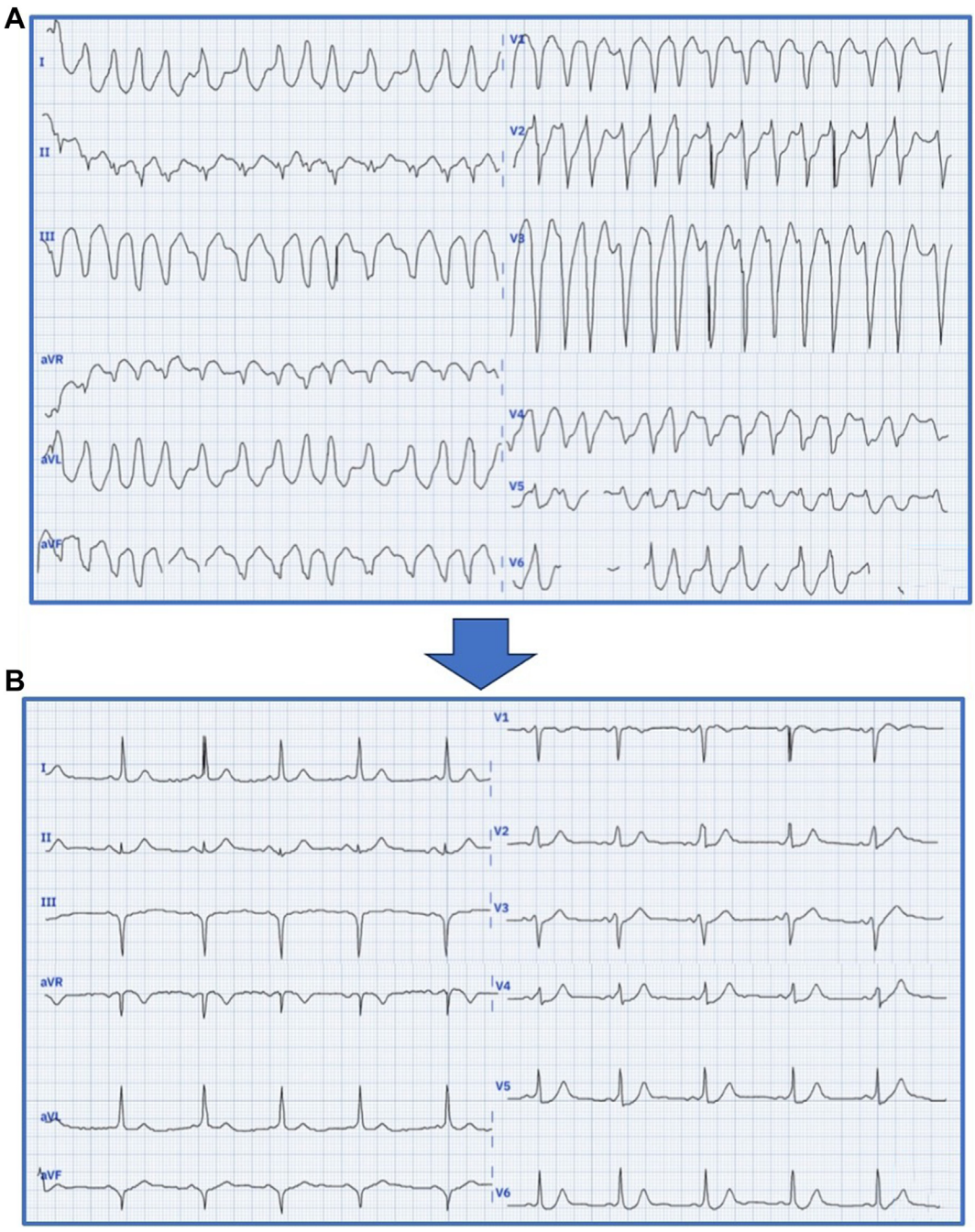

Figure 1. (**A**) Electrocardiogram findings at admission: accordion-like morphology with an irregularly irregular wide-complex tachycardia suggestive of pre-excited atrial fibrillation. (**B**) The electrocardiogram after electrical cardioversion, revealing a shortened PR interval and a negative delta wave in V1, along with a QRS transition occurring before V3 and with the most prominent positive delta wave observed in AVL, which suggests the presence of a right posteroseptal atriofascicular tract.

rapid ventricular response, which can lead to hemodynamic instability and adverse outcomes.[3] The complex anatomic features associated with Ebstein anomaly, such as a significantly enlarged right heart, a displaced tricuspid annulus, and distorted anatomic landmarks, can pose increased difficulties during catheter ablation procedures. Achieving and maintaining catheter stability can be challenging due to these factors, as experienced in our case. The surgical intervention chosen plays a pivotal role in the management of Ebstein anomaly among adult patients. Tricuspid valve repair is frequently favoured, 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 in which severe RV dysfunction is present.[6] Customized treatment strategies, interdisciplinary care, and regular long-term monitoring are crucial in optimizing outcomes for adult patients with Ebstein anomaly who undergo surgical treatment. In our case, the

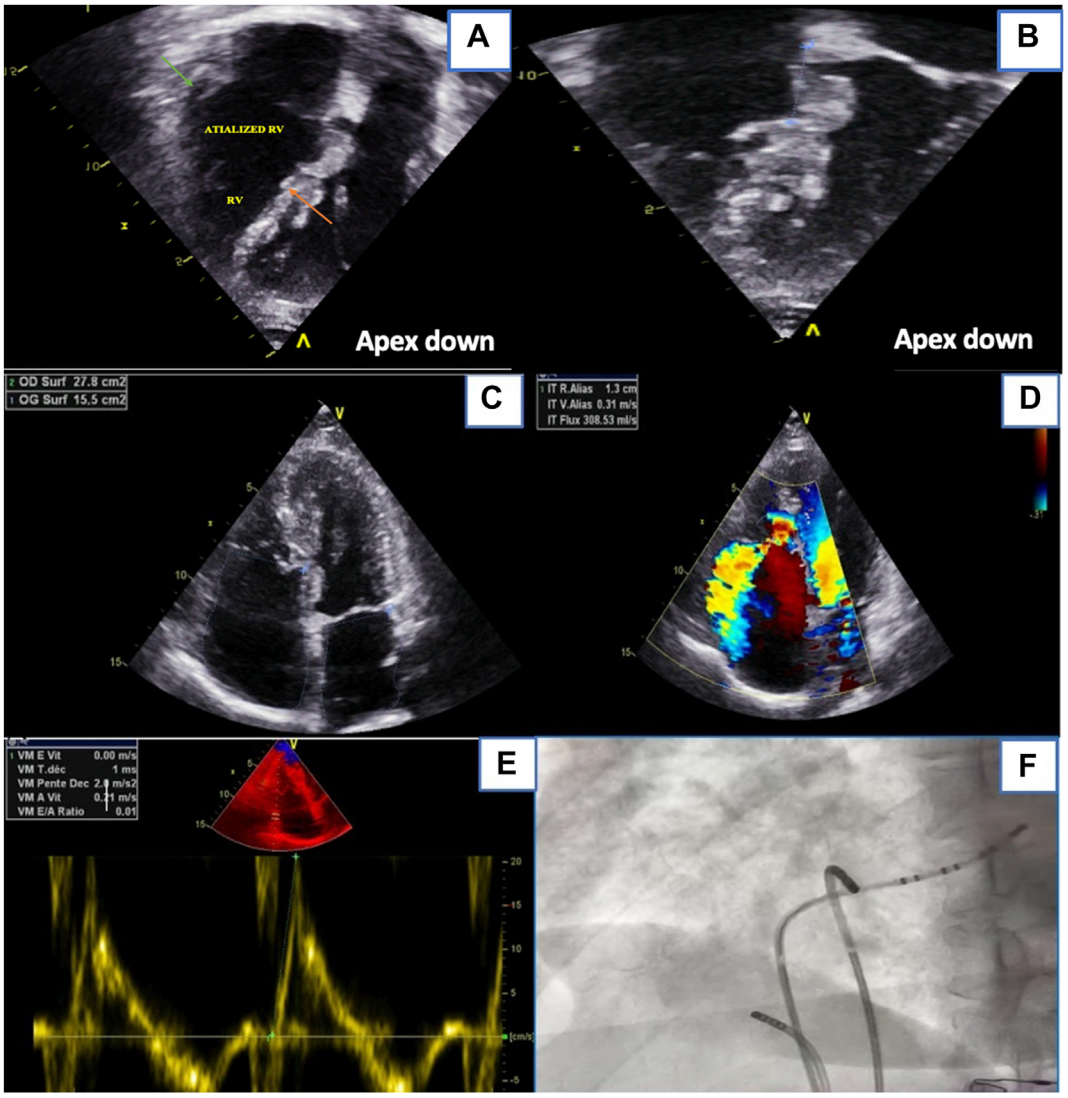

Figure 2. (**A, B**) Apical 4-chamber view reveals significant apical displacement of the septal tricuspid leaflet (**orange arrow**) relative to the anterior mitral leaflet (**green arrow**), leading to atrialization of the right ventricle. (**C**) Apical 4-chamber view demonstrates pronounced enlargement of the right atrium. (**D**) Color-flow Doppler shows evidence of severe tricuspid valve regurgitation. (**E**) Longitudinal systolic function of the right ventricle appears normal based on S′ wave and isovolumic acceleration. (**F**) Position of catheters during an electrophysiology study. We note the massively enlarged right heart with the displaced tricuspid annulus and the distortion of anatomic landmarks, making the procedure difficult.

main issue pertaining to Ebstein anomaly was related to rhythmic abnormalities. Therefore, following a multidisciplinary discussion within the heart team, and considering the patient's preference, the decision was made to put the patient on medical treatment.

Conclusion

This case report highlights the importance of considering rare cardiac conditions, such as Ebstein anomaly, even in individuals of advanced age. The coexistence of atrial fibrillation with pre-excitation 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.

Ethics Approval

The research reported had adhered to the relevant ethical guidelines.

Patient Consent

Written informed consent was obtained from the patient for publication of this study and accompanying images.

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Disclosures

The authors have no conflicts of interest to disclose.

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Supplementary Material

To access the supplementary material accompanying this article, visit *CJC Open* at https://www.cjcopen.ca/ and at https://doi.org/10.1016/j.cjco.2023.12.009.