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Radiofrequency Ablation of Atrial Flutter in 59-Year-Old Patient with Uncorrected Ebstein's Anomaly

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Abstract **Case Report**

Ebstein's anomaly (EA) is an uncommon congenital heart disease. Characterized by abnormal formed and apical displaced leaflets of the TV. The apical displacement of the tricuspid valve (TV) anatomically manifested by a morphological right atrium (RA), an atrialized segment of the right ventricle (RV), and the remaining functional RV. The TV is often regurgitant, resulting in right atrial and RV enlargement. The dilated right atrium in EA creates a fertile environment for atrial arrhythmias requiring radiofrequency ablation. Specific issues make the procedure more challenging in those patients. We report a case of successful radiofrequency ablation of cavo tricuspid isthmus-dependent atrial flutter in a patient of 59-year-old, with uncorrected Ebstein's anomaly.

Keywords: Ebstein's anomaly, Atrial flutter, Radiofrequency ablation, adults congenital heart disease.

INTRODUCTION

Ebstein's anomaly (EA) is a rare congenital heart condition, constituting less than 1% of all congenital cardiac malformations. Characterized by abnormal formed and apical displaced leaflets of the TV. The presentation ranges from a severe symptomatic form in the neonatal period to incidental detection later in life, including late adulthood. Arrhythmias are the leading cause of symptoms in adolescents and adults [1], requiring radiofrequency ablation. Catheter ablation of the tricuspid isthmus in EA is challenging because of the anomalous anatomy of this region [1]. Here, we report a case of successful radiofrequency ablation of atrial flutter in a 59-year-old patient with uncorrected Ebstein's anomaly.

CASE PRESENTATION

A 59-year-old woman visited emergency department complaining of dizziness, dyspnea (NYHA III) and palpitations that began three days prior. She reports intermittent palpitations for nearly two years. She was not taking any medications and had no significant family history of cardiac disease.

Upon admission, a 12-lead electrocardiogram was done and showed atrial flutter with 2:1 atrioventricular conduction (Figure 1). The patient was hemodynamically stable. The echocardiogram showed a dilated right ventricle with reduced longitudinal function and dilated right atrium, an apical displacement of the attachment of the septal tricuspid valve leaflet by 16 mm (9,6 mm/m²) compared to the attachment of the anterior mitral valve leaflet, with severe tricuspid regurgitation without stenosis, consistent with Ebstein's anomaly (Figure 2&3). There were no other valvular defects, and the left ventricular systolic function and size were normal (EF: 62%). The biological assessment showed no abnormalities.

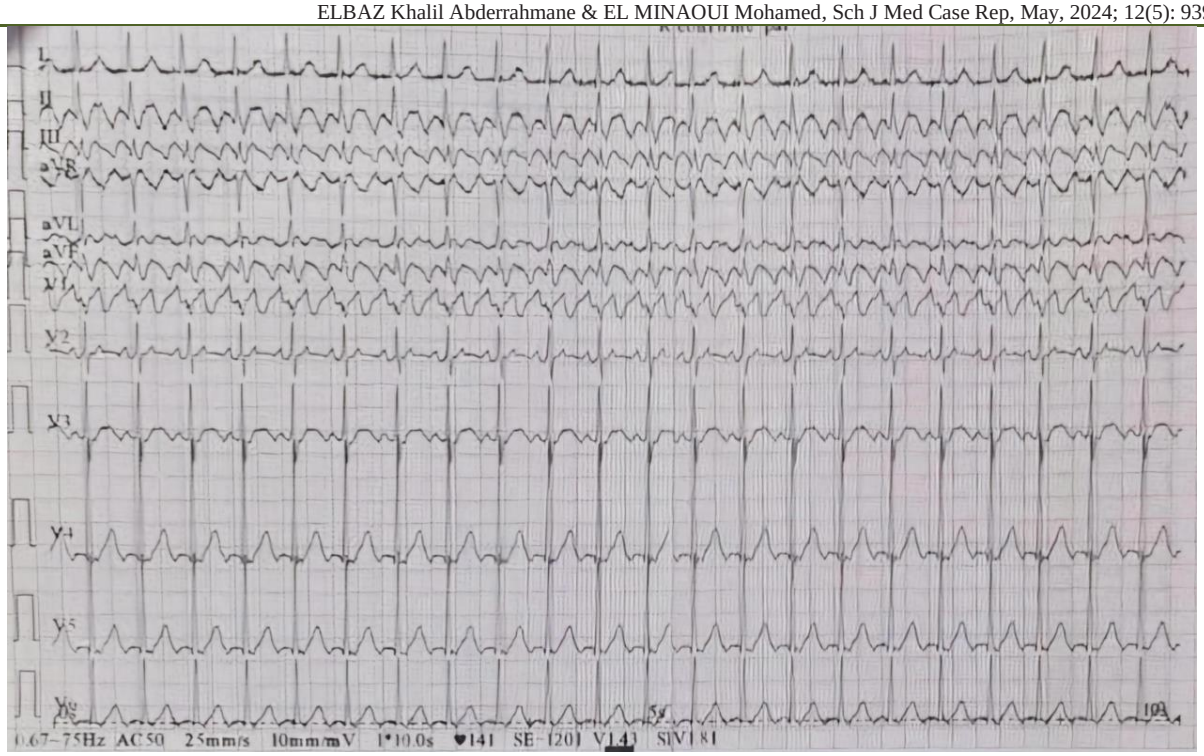

Figure 1: 12-lead electrocardiogram was done and showed atrial flutter with 2:1 atrioventricular conduction

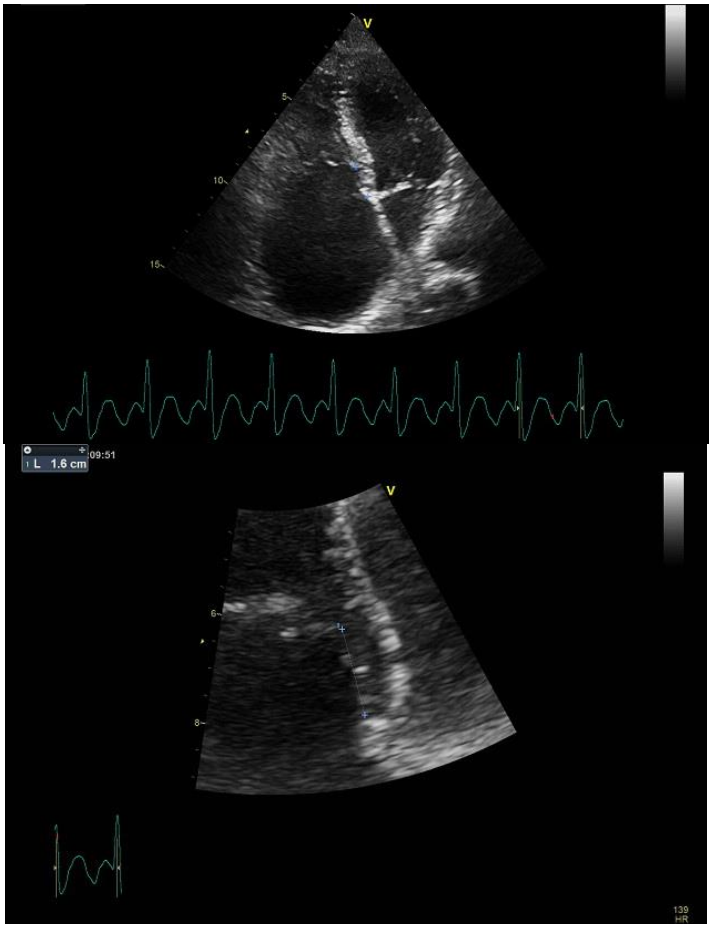

Figure 2&3: The dilation of the right heart cavities and the apical displacement of the tricuspid leaflets

The management includes rate control using betablockers, and anticoagulation therapy. She was referred for an electrophysiology study. The procedure was guided by fluoroscopy, investigation is conducted by

means of a double puncture of the right femoral vein and the subsequent fitting of two leads. One of these is a hexapolar lead, which is placed in the coronary sinus, while the other is a decapolar lead, which is positioned on the lateral wall of the right atrium.

The primary atrial depolarization was observed in the proximal coronary sinus, which indicated that the flutter originated from the right atrium. This was accompanied by an anti-clockwise activation.

Radiofrequency current of 60W was delivered, the flutter ceased, and the atrial signal shifted (Figure 4), with the disappearance of the collision front and bidirectional block, indicating successful ablation of the Cavo tricuspid isthmus. The procedure was completed without any complications.

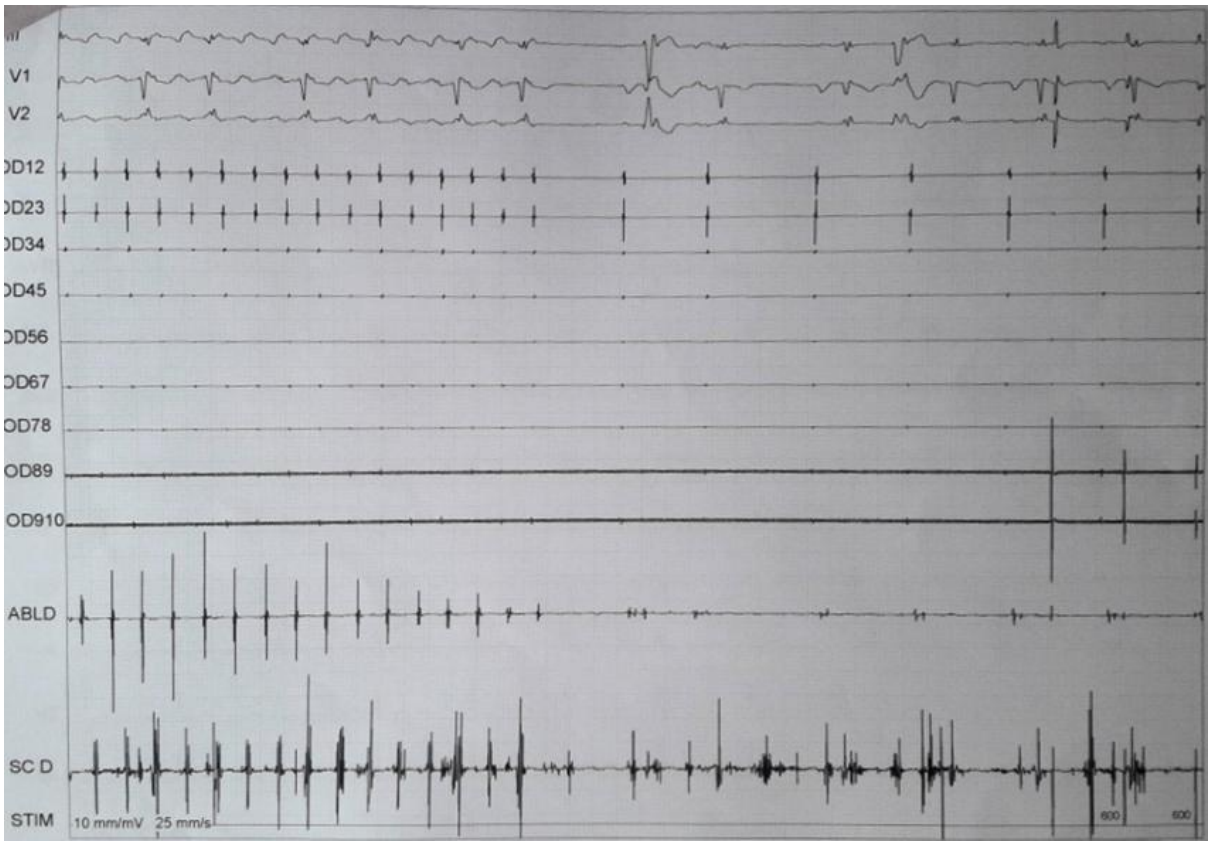

Figure 4: Intracardiac electrocardiogram, note the shift of atrial signal after Radiofrequency current

The patient left the electrophysiology lab in sinus rhythm, and at the completion of the procedure, the presenting atrial arrhythmias could not be reinduced.

The patient was followed up serially for two years and showed no recurrence of arrhythmias or symptoms.

DISCUSSION

Ebstein's anomaly (EA) is an uncommon congenital heart disease, Constituting less than 1% of all congenital cardiac malformations. First described on autopsy by Wilhelm Ebstein in 1866 [2]. EA characterized by apical displacement of the posterior and septal leaflets of the tricuspid valve (TV). The apical displacement of the TV anatomically manifested by a morphological right atrium (RA), an atrialized segment of the right ventricle (RV), and the remaining functional RV. The TV is often regurgitant, resulting in RA and RV enlargement, which provides the substrate for the development of both supraventricular and ventricular tachyarrhythmias [3].

Arrhythmias are the key symptoms in adolescents and adults, most frequently, atrioventricular reentry tachycardia, followed by intra-atrial re-entrant tachycardia (IART) in case of previous surgery, or cavo tricuspid isthmus-dependent atrial flutter (CTI-AFL).

However, the predominant arrhythmia in a cohort of 22 patients with EA was CTI-AFL [4]. In addition, CTI-AFL can be observed both after surgical incision of the right atrium or without previous surgery [5].

Catheter ablation using radiofrequency current attempting localized interruption and dissection circumscribed arrhythmogenic substrates has been now presented as a first-line therapy in international guidelines for such arrhythmias [6,7].

However, catheter ablation of CTI-AFL is considerably more challenging in EA patients compared to others, as evidenced by the frequent need for repeat procedures. The enlargement of the right-sided cavities complicates catheter stabilization in the atrioventricular groove. Additionally, constructing an ablation line across the cavotricuspid isthmus in EA patients is difficult due to the dysmorphic tricuspid valve and the atrialized right ventricle [1,9]. Nevertheless, effective control of these arrhythmias is important for optimal patient outcomes.

Ebstein's anomaly is associated with an increased incidence of heart failure and arrhythmias. Only 5% of patients with Ebstein's anomaly survive till the age of 50 without surgical correction [8]. Our patient's presentation at this age, without the need for surgical correction in his lifetime, was highly unusual, as he remained asymptomatic until one year ago. She also refused surgery and took the option of antiarrhythmic therapy, but she remained symptomatic. Radiofrequency ablation was the most suitable method for her treatment, as it is minimally invasive, does not require general anesthesia, and carries a low risk of complications. During follow-up of 2 years, the patient was without any symptoms and remain in sinus rhythm.

We reviewed the current literature on the use of radiofrequency ablation in patients with CTI-AFL and EA. Numerous reports detail the efficacy and long-term outcomes of radiofrequency catheter ablation for atrial arrhythmias in this group of patients [1,4].

Radiofrequency ablation was feasible, safe, and effective in our patient with EA. Additional periprocedural difficulties due to the anatomic anomaly did not occur. This case reports a follow-up of patient with uncorrected EA, who underwent a radiofrequency ablation of symptomatic atrial flutter with successful results. Further studies evaluating CTI-AFL radiofrequency ablation in this population are necessary, but our experience supports an interventional approach.

CONCLUSION

CTI-AFL in patients with EA may occur with or without prior surgery. Antiarrhythmic drugs often fail to control tachycardias or prevent recurrences, particularly over the long term. Radiofrequency catheter ablation appears to be an effective method for managing these arrhythmias. Although it is a complex procedure, it has shown very acceptable success rates and maintains a safety profile, especially in high-risk patients.

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