

CASE REPORT

Successful ablation of coexistent Mahaim tachycardia and right posterior accessory pathway in a patient with Ebstein's anomaly

Ebstein anomalisi olan çocukta Mahaim taşikardi ve sağ arka aksesuar yol birlikteliğinin başarılı ablasyonu

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Summary– The atriofascicular accessory pathway (AP), known as the Mahaim pathway, is a rare form of pre-excitation, comprising less than 3% of all APs. Mahaim AP is characterized by decremental, anterograde-only conduction, and antidromic tachycardia with left bundle branch morphology. Prevalence of Mahaim AP in Ebstein's anomaly is significantly high. In addition, combination of Wolff–Parkinson–White (WPW) syndrome and Mahaim AP in patients with Ebstein's anomaly has been reported. Presently described is the coexistence of Mahaim AP and manifest WPW syndrome in a patient with Ebstein's anomaly, who was successfully ablated without fluoroscopy.

Özet– Atriyofasiküler aksesuar yolak (AY) diğer deyimle Mahaim yolları preeksitasyonun nadir formu olup aksesuar yolların %3'den daha azını oluşturmaktadır. Mahaim aksesuar yolak dekremental, sadece önedoğru iletisi olan ve sol dal bloklu antidromik taşikardi ile kendini gösterir. Mahaim aksesuar yolağın Ebstein anomalisindeki prevalansı oldukça yüksektir. Ayrıca Wolf-Parkinson-White (WPW) sendromu ve Mahaim AY birlikteliği Ebstein anomalisinde gösterilmiştir. Bu yazıda, Ebstein anomalisi olan hastada WPW ve Mahaim AY birlikteliğinin floroskopisiz başarılı ablasyonunu sunuyoruz.

Coexistence of Mahaim AP with manifest Wolff–Parkinson–White (WPW) syndrome in patients with Ebstein's anomaly has been reported.^[1] Treatment modality of multiple APs depends upon clinical significance and risk assessment of these pathways. Radiofrequency (RF) ablation can be used safely and effectively for treatment of various types of tachycardias in patients with Ebstein's anomaly.^[2]

was no family history of syncope, sudden cardiac death, or arrhythmias. Physical examination was normal. Electrophysiological study was planned for risk stratification of WPW syndrome.

Abbreviations:

AP	Accessory pathway
LBBB	Left bundle branch block
RF	Radiofrequency
SVT	Supraventricular tachycardia
WPW	Wolff–Parkinson–White

CASE REPORT

A 12-year-old boy with a diagnosis of Ebstein's anomaly was referred due to palpitations. Twelve-lead resting electrocardiography revealed short PR interval (110 msec) and wide QRS duration (119 msec) with delta waves consistent with manifest WPW syndrome. No tachycardia was observed on electrocardiography. Echocardiogram revealed Ebstein's anomaly with normal right and left ventricular functions. There

Electrophysiological study was performed under general anesthesia without intubation. Quadripolar catheters were positioned at the high atrium, His bundle area, and right ventricular apex. A steerable decapolar catheter was inserted into the coronary sinus for positional reference. EnSite NavX system (St. Jude Medical, Inc., Little Canada, MN, USA) was used for 3-dimensional mapping and catheter navigation. Right atrial anatomy was reconstructed without fluoroscopy. Baseline electrophysiological measurements were as follows: AH interval of 32 msec, HV interval of 24

Received: August 23, 2015 Accepted: December 03, 2015

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msec, and baseline cycle length of 582 msec. There was evidence of pre-excitation consistent with right-sided AP. Wide QRS tachycardia with left bundle branch block (LBBB) and inferior axis pattern was induced spontaneously due to catheter manipulation. QRS morphology was different from baseline pre-excited QRS morphology (Figure 1). Atrial pacing demonstrated a difference in pre-excited QRS morphology, compared to tachycardia QRS morphology (Figure 2). Tachycardia cycle length was 280 msec, and was not considered antidromic supraventricular tachycardia (SVT) of the pathway leading to baseline pre-excitation. Tachycardia was terminated with overdrive pacing due to hemodynamic impairment during tachycardia.

To assess risk stratification of WPW, effective refractory period of AP and shortest pre-excited RR duration during atrial fibrillation were measured (effective refractory period was 320 msec and shortest pre-excited RR was 338 msec). Morphology of tachycardia during atrial fibrillation was different from baseline QRS morphology. However, this tachycardia was similar to the wide QRS tachycardia previously induced. Due to unchanged QRS morphology in spite of irregularity in RR intervals, this wide QRS tachycardia was not interpreted as aberrant SVT, but rather as Mahaim tachycardia. During the mapping around the tricuspid annulus, typical Mahaim potential was detected at the lateral tricuspid annulus (Figure 3).

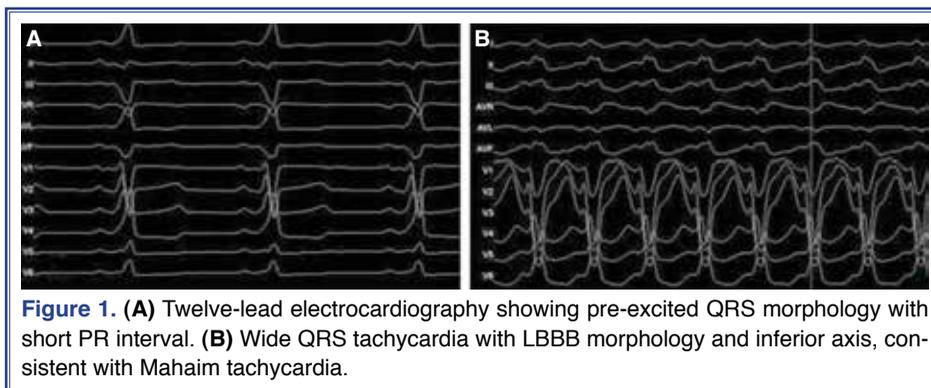


Figure 1. (A) Twelve-lead electrocardiography showing pre-excited QRS morphology with short PR interval. (B) Wide QRS tachycardia with LBBB morphology and inferior axis, consistent with Mahaim tachycardia.

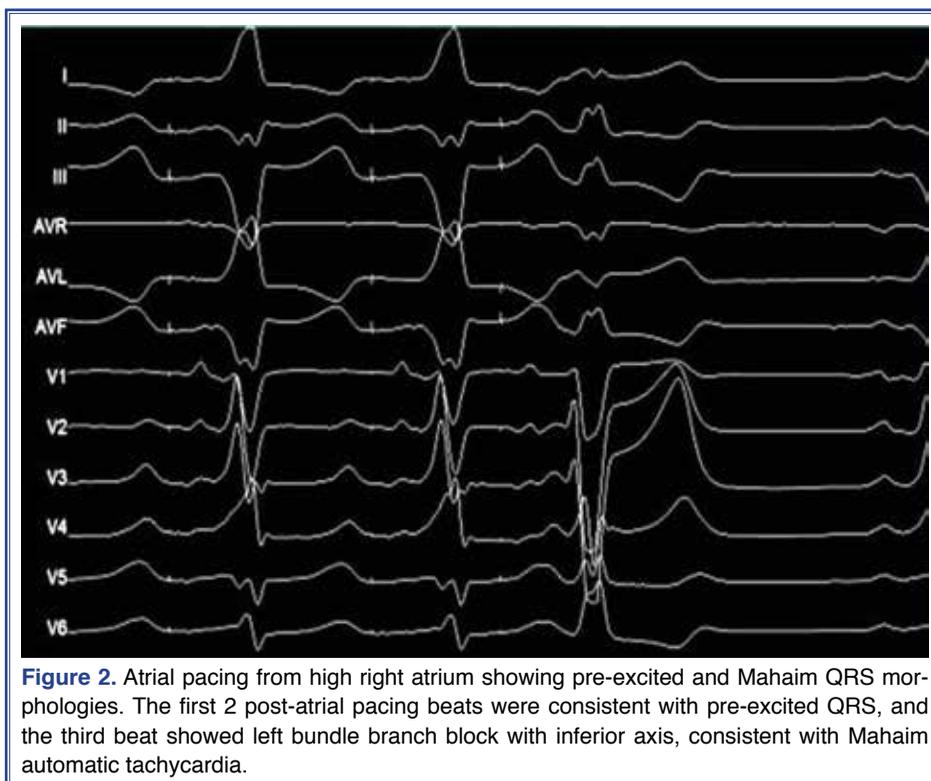


Figure 2. Atrial pacing from high right atrium showing pre-excited and Mahaim QRS morphologies. The first 2 post-atrial pacing beats were consistent with pre-excited QRS, and the third beat showed left bundle branch block with inferior axis, consistent with Mahaim automatic tachycardia.

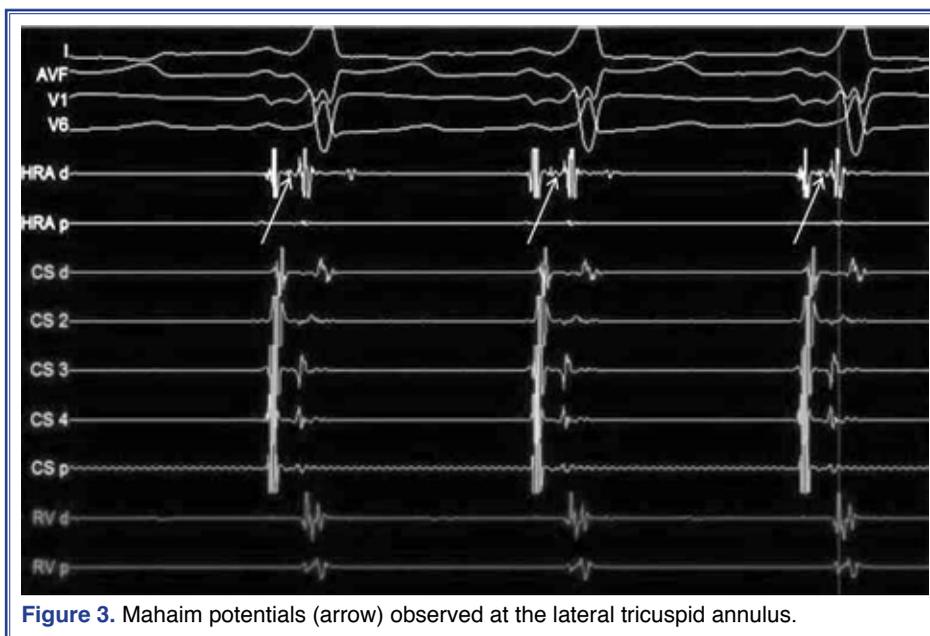


Figure 3. Mahaim potentials (arrow) observed at the lateral tricuspid annulus.

RF energy was delivered to this region and an accelerated wide QRS rhythm with similar QRS morphology to Mahaim tachycardia was detected during ablation. This finding also supported evidence of Mahaim AP. Following successful RF and during atrial pacing, the wide QRS morphology of the Mahaim pathway was not observed again, though pre-excitation was still noted at baseline. Antegrade and retrograde mapping located the pathway in the right posterior area of the tricuspid valve annulus. AP was eliminated at the 14th second of RF ablation. Following successful ablation and during ventricular pacing, no retrograde conduction was observed. At the end of the post-procedural waiting period of 30 minutes, atrioventricular block was obtained with adenosine administration, and no pre-excitation was observed. It was conclusively confirmed that the antegrade limb of the wide QRS tachycardia was Mahaim tachycardia, using the right posterior AP as the retrograde limb, as no retrograde conduction was present at the atrioventricular node. Total procedure time was 230 min, and no fluoroscopy was used.

DISCUSSION

The most accepted and successful site of catheter ablation of Mahaim pathways is at the site of Mahaim potential,^[3,4] though ablation at the septal insertion site near the right bundle branch can be performed. Mahaim pathway is characterized by antegrade

conduction and decremental conduction property.^[4] Tachycardia shows LBBB morphology, with left axis deviation.^[4] Location of the pathway is at the lateral tricuspid annulus in most cases.^[5] Mahaim automatic tachycardia occurs during RF ablation due to heat-related automaticity and has been considered a marker of successful ablation.^[6] Incidence of multiple APs in patients with Ebstein's anomaly is high (50%).^[11] In addition, the combination of a Kent and Mahaim-like AP has been reported.^[7,8] In cases of antidromic tachycardia, the antegrade limb of the pathway is Mahaim AP, and the retrograde limb of the reentrant circuit may be APs or atrioventricular node.

Typical Mahaim tachycardia has LBBB and superior axis. In the present case, so-called Mahaim tachycardia had an inferior axis. However, the localization and insertion site of the Mahaim AP may be very bizarre, including the anterior side of the right and left ventricles, known as atypical Mahaim conduction. Although the Mahaim AP may follow a pathway that is located and may be ablated at the lateral tricuspid annulus, the insertion site is generally in inferior septal or posterior regions, as in the present case. Differential diagnoses for wide complex tachycardia include SVT with aberrancy, ventricular tachycardia, and antidromic tachycardia. In the present case, 2 diagnoses of wide QRS tachycardia were possible: aberrant SVT and Mahaim tachycardia. Therefore, the differentiation between these types of tachycardia should be discussed.

Several findings support the diagnosis of Mahaim AP:

1. The presence of Mahaim potentials in the lateral annulus, a weak indicator of Mahaim AP.
2. Accelerated rhythm from the Mahaim pathway during RF ablation, the strongest indicator.
3. QRS morphology that does not change, in spite of irregularity in the RR intervals during atrial fibrillation. The present morphology was consistent with Mahaim antegrade conduction, rather than aberrant conduction.
4. Two wide QRS morphologies observed during atrial pacing, one consistent with right posterior, another with Mahaim antegrade conduction.

RF ablation remains the treatment of choice for this disorder. However, treatment of Mahaim tachycardia with cryoablation has also been reported.^[9]

In conclusion, patients with Ebstein's anomaly can have multiple APs, including Mahaim pathways. A careful and thorough electrophysiological assessment is needed in order to successfully achieve ablation in these cases.

Conflict-of-interest issues regarding the authorship or article: None declared.

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Keywords: Accessory pathway; Ebstein's anomaly; Mahaim pathway.

Anahtar sözcükler: Aksesuar yolak; Ebstein anomalisi; Mahaim yolu.