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A Rare Case of Wolff–Parkinson–White Syndrome Associated with Right Atrial Aneurysm

Sağ Atriyal Anevrizma ile İlişkili Nadir Bir Wolff-Parkinson-White Sendromu Olgusu

ABSTRACT

Wolff–Parkinson–White syndrome is rarely associated with a right atrial aneurysm. However, when such a condition occurs, it will be hard to manage since pre-excitation will be induced as long as the aneurysm persists. A 14-year-old female patient received emergency treatment for irregular wide QRS complex tachycardia in our center, and a pre-excitation pattern was then observed on the surface electrocardiogram. An initial electrophysiological study revealed a high-risk right posterior accessory pathway that was resistant to both radiofrequency and irrigated radiofrequency ablations. Subsequently, fluoroscopy showed that this was due to a right atrial aneurysm. Although successful ablation with irrigated radiofrequency was performed in the second procedure, the procedure was considered suboptimal due to the association of aneurysm. Accordingly, we initiated anti-thrombotic and anti-arrhythmic drug therapy. We decided to omit surgery and followed the case under medical treatment for 2 years without complications. Here, we report this rare co-existence and our treatment approach in detail.

Keywords: Children, right atrial aneurysm, Wolff–Parkinson–White syndrome

ÖZET

Wolff-Parkinson-White sendromunun sağ atriyal anevrizma ile ilişkili olarak görülmesi nadirdir. Ancak bu birliktelik durumunda, anevrizma devam ettiği sürece pre-eksitasyon indükleneceğinden yönetimin zor olacağı bilinmelidir. 14 yaşında kız hasta düzensiz geniş QRS kompleksli taşikardi nedeniyle hastanemiz acil servisine başvurdu. Yüzey elektrokardiyogramında pre-eksitasyon paterni gözlenmesi üzerine tarafımıza danışılan hastanın ilk elektrofizyolojik çalışmasında, hem radyofrekans hem de irrigasyonlu radyofrekans ablasyonlarına dirençli, yüksek riskli bir sağ posterior aksesuar yol görüldü. Floroskopide, bu aksesuar yolun sağ atriyal anevrizmaya bağlı olduğunu görüldü. İkinci işlemde, irrige RF ile başarılı ablasyon yapılmasına rağmen, anevrizma birlikteliği nedeniyle prosedür yetersiz kabul edildi. Bu nedenle antitrombotik ve antiaritmik ilaç tedavisine başlandı. Hasta, cerrahi düşünülmeksizin, 2 yıldır medikal tedavi altında sorunsuz takip edilmektedir. Burada nadir görülen bu birlikteliği ve tedavi yaklaşımımızı detaylı olarak sunuyoruz.

Anahtar Kelimeler: Çocuk, sağ atrial anevrizma, Wolff-Parkinson-White sendromu

Wolff–Parkinson–White (WPW) syndrome is a congenital rhythm disorder characterized by pre-excitation due to rapid conduction through the accessory atrioventricular pathway.

The electrocardiographic (ECG) finding of the WPW pattern consists of a short PR interval and prolonged QRS with an initial slurring upstroke ("delta" wave) in the presence of sinus rhythm. The diagnosis of WPW syndrome requires a combination of the mentioned ECG finding, tachyarrhythmia, and clinical symptoms of tachycardia, such as palpitations, episodic dizziness, presyncope, syncope, and even cardiac arrest. Despite occurring in only 0.1%-0.3% of the cases, the WPW pattern, that is, pre-excitation, is critical since it can cause life-threatening arrhythmias. Wolff–Parkinson–White syndrome is commonly seen in children with normal hearts, but its incidence increases in the presence of Ebstein anomaly and hypertrophic cardiomyopathy.[1] Wolff–Parkinson–White syndrome associated with an atrial aneurysm is remarkably less frequent.[2]

CASE REPORT
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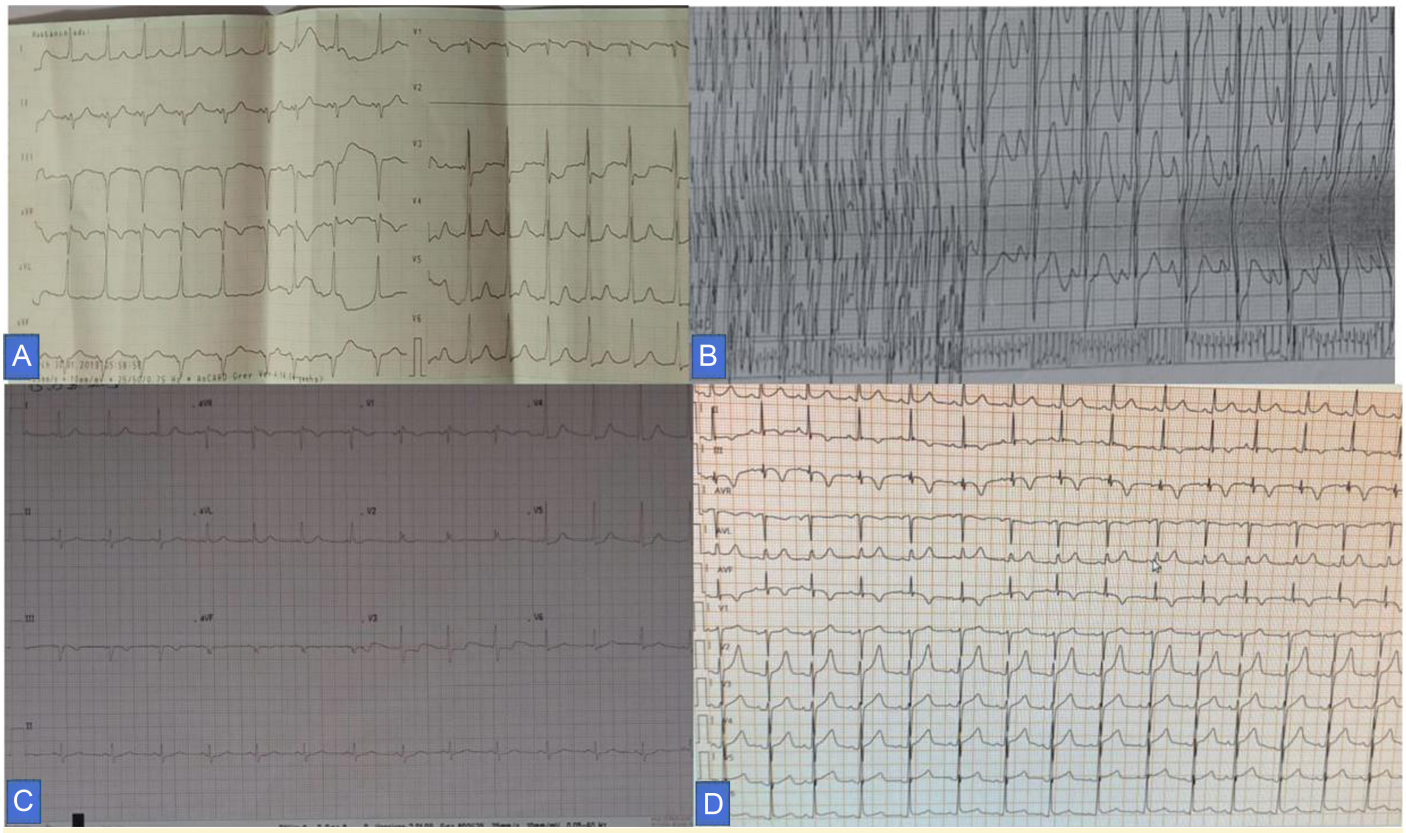

Figure 1. Electrocardiographic images (findings). (A) Surface electrocardiography (ECG) before the first ablation; Delta wave and QRS polarities indicate the presence of right postero-septal accessory pathway according to modified Arruda classification. (B) Fast and sustained supraventricular tachycardia attack was seen during 24 hours Holter at the first admission. (C) Surface ECG after the first ablation. Although the degree of pre-excitation has decreased compared to the pre-procedure, there are still signs of pre-excitation. (D) Surface ECG after the second ablation; manifest pre-excitation was not observed.

Right atrial aneurysm is a rare anomaly of unknown origin. Approximately half of the patients with right atrial aneurysms have normal sinus rhythm, but about 28% have atrial fibrillation and flutter. In some cases, pre-excitation syndrome, junctional rhythm, heart block, and supraventricular tachycardia have also been noted.[3] Here, we report a rare co-existence of WPW syndrome and right atrial aneurysm.

Case Report

A 14-year-old girl was referred to the emergency room of our hospital due to syncope occurring 3 times on the same day. The patient was hemodynamically stable and her ECG revealed an irregular wide QRS complex tachycardia thought to be atrial fibrillation with rapid conduction through the accessory pathway (AP). The condition was successfully treated with cardioversion, and the patient was transferred to the cardiology department. There, a WPW pattern was observed on the surface ECG and a fast and sustained supraventricular tachycardia (SVT) attack was seen during the 24-hour Holter monitoring (Figure 1A and 1B). Additionally, mitral valve prolapse and mild mitral regurgitation were detected in the subsequent echocardiographic evaluation. Subsequently, pre-excitation persisted at the highest rate of exercise stress testing which suggests an AP may present a high risk of sudden cardiac death. Then, an electrophysiological study uncovered a high-risk right posterior AP. During the 3D En Site mapping, the aneurysm structure and ablation points under the coronary sinus ostium in the antero-posterior and postero-anterior positions were remarkable (Figure 2A and 2B). A concentric sequence in the coronary sinus during orthodromic SVT was evidence of the accessory pathway in the right atrium (Figure 2C). Also, we saw the transient bump effect as a result of the mechanical consequence of the catheter tip in the aneurysm area (Figure 2D). Atrial fibrillation occurred spontaneously during the first procedure. The shortest pre-excited RR distance at this time was 200 milliseconds. The accessory pathway effective refractor period was 210 milliseconds. Finally, VA conduction terminated at 200 milliseconds and was easily inducible in orthodromic SVT. All these findings further confirmed the high risk of the pathway. However, both radiofrequency and irrigated radiofrequency ablations were ineffective due to a right atrial aneurysm determined by fluoroscopy during the procedure. Figure 1C shows the surface ECG after the first ablation. We noticed that the

ABBREVIATIONS

AP	Accessory pathway
ECG	Electrocardiography
MRI	Magnetic resonance imaging
SVT	Supraventricular tachycardia
WPW	Wolff–Parkinson–White

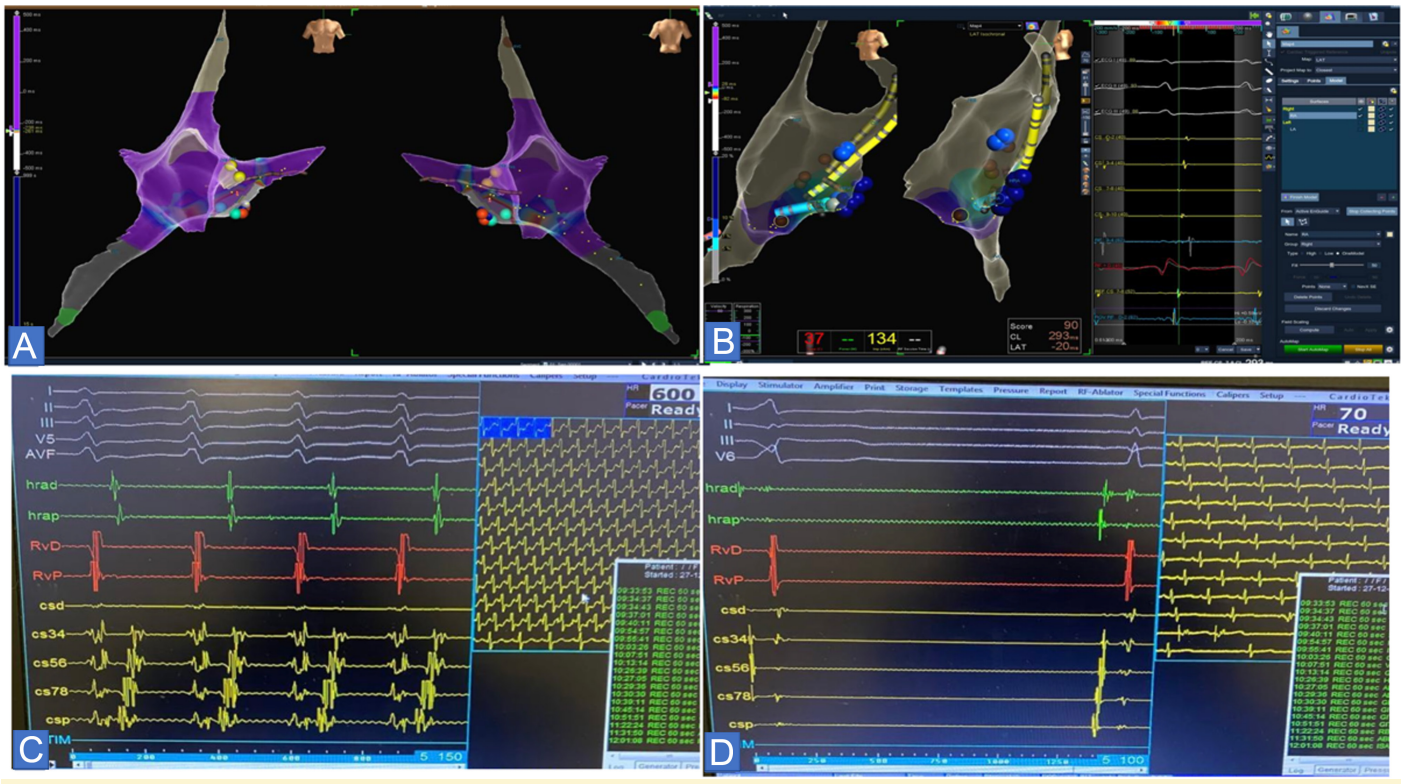

Figure 2. Electrophysiological images (findings). (A) During the 3D En Site mapping, the aneurysm structure and ablation points under the coronary sinus ostium in the antero-posterior and postero-anterior positions are remarkable. (B) LAO and left lateral view of the same aneurysm. (C) Concentric sequence in the coronary sinus during orthodromic sustained supraventricular tachycardia is evidence of the accessory pathway in the right atrium. (D) Transient bump effect as a result of the mechanical effect of the catheter tip in the aneurysm area.

accessory pathway localization was identical to the aneurysm localization because we obtained AP signals with ablation catheters in the atrial aneurysm images in fluoroscopy. Magnetic resonance imaging (MRI) was performed to define better the location and size of the aneurysm (Figure 3). The patient was discharged on oral sotalol therapy. Approximately a year later, irrigated radiofrequency ablation was repeated, and the AP was successfully eliminated (Figure 1D). However, the procedural outcome was considered sub-optimal since the AP had an epicardial connection associated with the right atrial aneurysm. It was then determined that the high-risk AP had already turned into a low-risk one after the first procedure. We decided to continue the sotalol therapy following the catheter ablation, assuming that the aneurysm may be involved in the persistent pre-excitation. Besides, acetylsalicylic acid therapy was started to prevent thromboembolic complications. No complications regarding radiofrequency catheter ablation and tachyarrhythmia occurred during the 2-year follow-up.

Written informed consent was obtained from the patients' legal guardians.

Discussion

The approach to WPW syndrome varies depending on the patient's age, weight, and symptoms. Symptoms accompanied by a shortest pre-excited R-R interval (SPERRI) during atrial fibrillation <250 milliseconds are known to increase the risk of sudden death.[4] However, recent pediatric studies have shown that nonpersistent pre-excitation cases may still pose a high risk of life-threatening arrhythmias.[5] Exercise test is important for

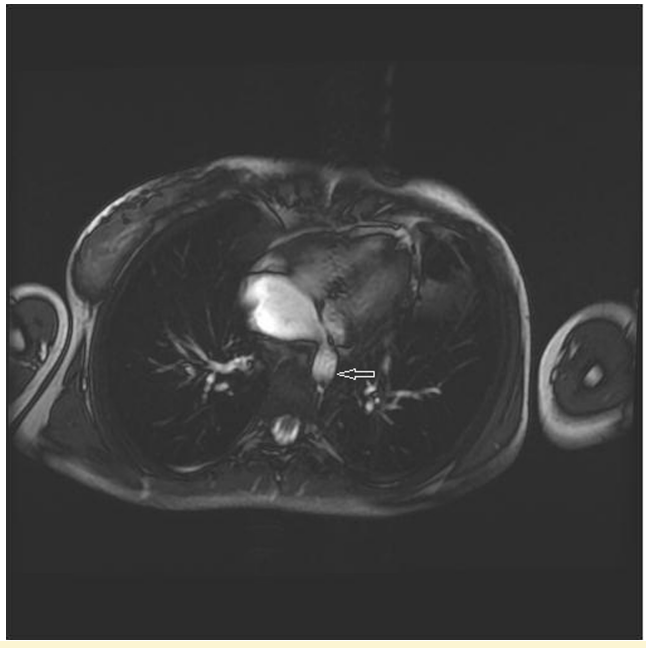

Figure 3. Arrow: Right atrial aneurysm on magnetic resonance imaging.

risk classification. At the faster heart rates of exercise stress testing, the presence of an abrupt and clear loss of pre-excitation suggests a weak or low-risk pathway. On the other hand, the persistence of pre-excitation during the entire ambulatory monitoring period suggests that an invasive evaluation may be further needed. However, it does not necessarily mean that the pathway is "high-risk".[2] As a pre-excitation pattern existed in our case at the faster heart rate of exercise stress testing, we performed an electrophysiological study.

Invasive electrophysiological study and radiofrequency ablation are recommended for children with body weight >15 kg and age >5 years when complications risk is low and medical center experience in pediatric cases is high.[6] Our case had a SPERRI <200 milliseconds and qualified as high risk. Considering the patient's life-threatening arrhythmia and symptoms, we decided to proceed with catheter ablation.

Most right atrial aneurysm cases manifest no symptoms. So, the condition may be diagnosed incidentally by detecting cardiomegaly or abnormal shadow on the chest x-ray. Symptomatic patients usually experience atrial arrhythmias. However, it should be kept in mind that a right atrial aneurysm may occasionally be associated with WPW syndrome. Patients diagnosed with right atrial aneurysm must be evaluated for other arrhythmias, such as junctional rhythm, atrioventricular block, unremitting supraventricular tachycardia, and atrial fibrillation. The increased likelihood of pre-excitation and atrial fibrillation considerably elevates the risk of sudden cardiac death. Additionally, the patient should receive anticoagulants to prevent thrombus formation.[7]

The management of right atrial aneurysms is still controversial. According to the literature, less than half of all cases are referred to surgery. Surgical treatment can yield satisfactory outcomes and reduce the incidence of life-threatening complications such as persistent arrhythmias.[8] Movsesyan et al[9] reported a case with a right atrial aneurysm associated with WPW syndrome which underwent surgery through the complete median sternotomy approach. Likewise, Mirhosseini et al[10] reported treating a giant right atrial aneurysm surgically. They concluded that while small asymptomatic aneurysms can be treated conservatively, surgery is reserved for large symptomatic aneurysms. Antithrombotic and antiarrhythmic drug therapy should be considered for asymptomatic patients without a giant right atrial aneurysm.[11]

The high-risk AP in our case had turned into a low-risk one, and we could regularly measure the aneurysm size thanks to patient compliance, so she has been followed up with sotalol and acetylsalicylic acid therapy.

Informed Consent: Written informed consent was obtained from the patients' legal guardians.

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