CASE REPORT

Aneurysm of the muscular septum associated with Wolf-Parkinson-White syndrome presenting as dilated cardiomyopathy; A report of two cases

Dilate kardiyomiyopati ile ortaya çıkan olan Wolf-Parkinson-White sendromu ile ilişkili musküler septal anevrizma; İki olgu sunumu

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Summary– Muscular septal aneurysms are extremely rare without a ventricular septal defect and are diagnosed accidentally in most cases. Reported cases generally have rhythm disturbance or electrocardiographic findings of Wolf-Parkinson-White (WPW) syndrome. Presently described are 2 cases of ventricular septal aneurysm associated with WPW syndrome, which presented as dilated cardiomyopathy. Pre-excitation disappeared gradually in first patient. There was also concurrent decrease in degree of bulging of the interventricular septum and improvement of left ventricular systolic function. Second patient had complaints of palpitation and was referred for ablation of accessory pathway. Our findings suggest that presence of pre-excitation may lead to ventricular dyssynchrony and abnormal ventricular septal movement, resulting in appearance of aneurysm.

Özet– Musküler septal anevrizma ventriküler septal defektin eşlik etmediği durumlarda oldukça nadirdir ve olguların büyük kısmında rastlantısal olarak saptanır. Bildirilen olguların genellikle ritm problemi veya Wolf-Parkinson-White (WPW) sendromunun elektrokardiyografik bulguları vardır. Bu yazıda, dilate kardiyomiyopati şeklinde ortaya çıkan ve WPW sendromu ile ilişkili septal anevrizma saptanan iki olgu sunuldu. Olgulardan birinde preeksitasyon zamanla kaybolurken aynı zamanda interventriküler septumun deviasyonunda azalma ve sol ventrikül sistolik fonksiyonunda düzelme izlendi. İkinci olgu çarpıntı yakınmasının olması nedeniyle aksesuvar yolağın ablasyonu amacıyla kurumumuza yönlendirilmişti. Bulgularımız pre-eksitasyon varlığının ventrikül senkronizasyon bozukluğuna ve anormal ventrikül septal hareketine neden olarak anevrizma görüntüsünü oluşturduğunu düşündürmüştür.

Muscular septal aneurysms are extremely rare without a ventricular septal defect. It may be congenital or acquired.[1] Familial cases have been described.[2] Reported cases generally have rhythm disturbance or electrocardiographic findings of Wolf-Parkinson-White (WPW) syndrome.[3,4]

Abbreviations:

ACE	*Angiotensin converting enzyme*
CTR	*Cardiothoracic ratio*
DCM	*Dilated cardiomyopathy*
EF	*Ejection fraction*
FS	*Fractional shortening*
LV	*Left ventricle*
RV	*Right ventricle*
WPW	*Wolf-Parkinson-White*

A subset of patients with WPW syndrome may develop dilated cardiomyopathy (DCM). Although DCM may occur in symptomatic WPW patients with sustained tachyarrhythmia, recent observations suggest that significant left ventricular (LV) dysfunction may also arise in the absence of incessant tachyarrhythmia.[5–8]

Presently described are 2 cases of muscular ventricular septal aneurysm associated with WPW syndrome presenting as DCM. Written, informed consent was obtained from parents of both patients included in the report.

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CASE REPORT

Case 1– A 16-month-old boy was referred for precise evaluation after diagnosis of DCM. His physical examination was normal, except for grade 2/6 systolic murmur, best heard at left sternal border. Electrocardiogram showed large QRS complex and short PR interval and delta wave, features of WPW syndrome (Figure 1a). A 24-hour Holter electrocardiogram revealed intermittent WPW without any documented supraventricular tachycardia. Chest radiography indicated cardiomegaly with cardiothoracic ratio (CTR) of 0.64. Transthoracic echocardiography displayed bulging of muscular septum into right ventricle (RV) during systole with ejection fraction (EF) and fractional shortening (FS) of 40% and 19%, respectively (Figure 1b). Upon cardiac catheterization, LV angiogram showed muscular septal aneurysm bulging into RV (Figure 1c). Angiotensin converting enzyme (ACE) inhibitor and propranolol were initiated and continued without any significant complaint. On follow-up, gradual disappearance of WPW findings was observed on surface and 24-hour Holter electrocardiogram, and degree of bulging of septal aneurysm also decreased significantly. At 6 years of age, echocardiography indicated normal EF and FS, without indication of septal aneurysm and only rare, isolated, premature atrial contraction without pre-excitation on 24-hour Holter electrocardiogram.

Case 2– A 9-year-old immigrant girl was referred to our hospital with diagnosis of DCM. She was taking digoxin, furosemide, ACE inhibitor, and carvedilol on admission. She complained of palpitations without documented supraventricular or ventricular tachycardia. Physical examination revealed normal

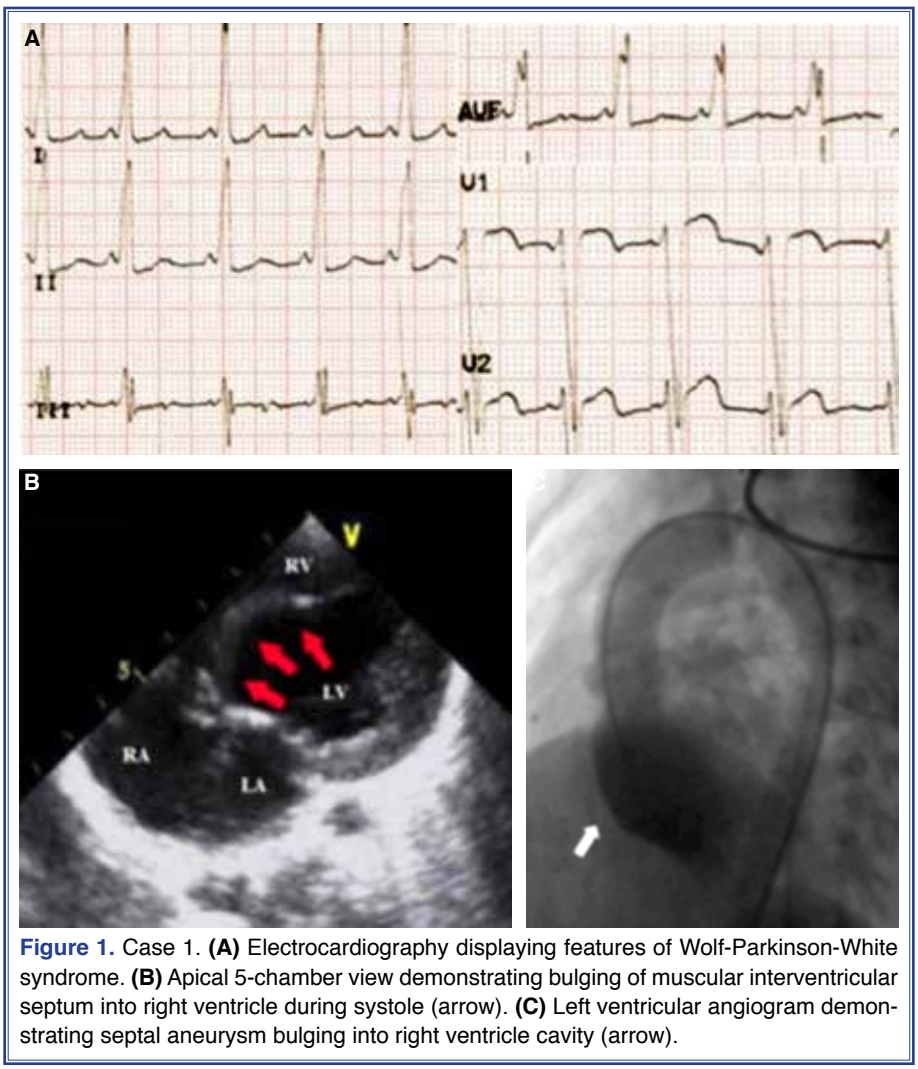

Figure 1. Case 1. **(A)** Electrocardiography displaying features of Wolf-Parkinson-White syndrome. **(B)** Apical 5-chamber view demonstrating bulging of muscular interventricular septum into right ventricle during systole (arrow). **(C)** Left ventricular angiogram demonstrating septal aneurysm bulging into right ventricle cavity (arrow).

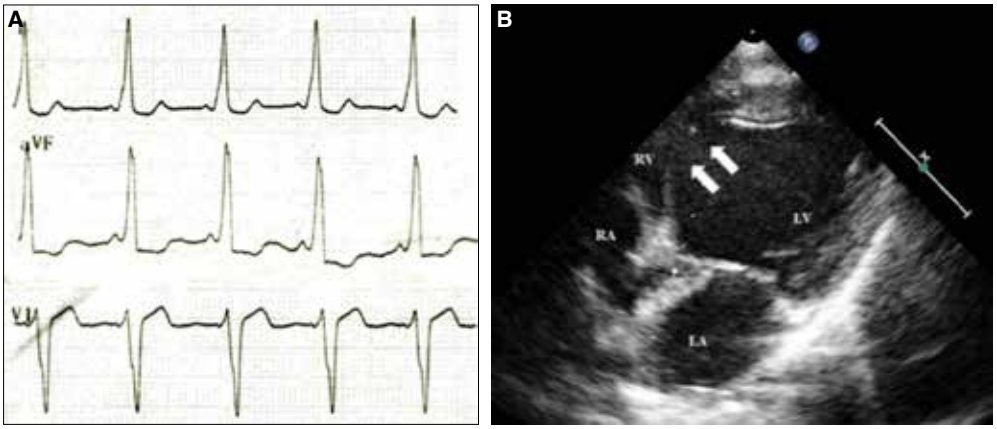

Figure 2. Case 2. **(A)** Electrocardiography displaying features of Wolf-Parkinson-White syndrome. **(B)** Apical 4-chamber view demonstrating bulging of basal interventricular septum into right ventricle during systole (arrow).

findings except for grade 1/6 systolic murmur, best heard at left sternal border. A 12-lead electrocardiogram demonstrated pre-excitation findings, displaying characteristics of anteroseptal accessory pathway and pathological ST depression (Figure 2a). CTR was determined to be 0.54 on chest X-ray. Echocardiography revealed aneurysm of muscular septum bulging into RV during systole, with EF and FS of 46% and 22%, respectively (Figure 2b). Patient was diagnosed with muscular septal aneurysm with WPW syndrome and referred to another center for ablation of accessory pathway for recurrent palpitations.

DISCUSSION

Aneurysm of membranous septum often occurs as consequence of spontaneous closure of ventricular septal defect. Aneurysm of muscular septum, however, is extremely rare. Possible explanations for formation of these aneurysms include weakness of the septum due to genetic defect of mesenchymal cell migration from developing interventricular septum or abnormal ventricular septal movement due to rhythm disturbance.[2,3] Almost half of patients with muscular septal aneurysm are asymptomatic, and most cases are diagnosed incidentally.[1–4] Both of our patients were referred for evaluation of DCM. Since familial cases of muscular septal aneurysm have been reported, family screening was carried out in both cases, but no additional case was detected.

Muscular septal aneurysm associated with WPW has been reported previously.[3] However, causal relationship is not clear. It has been hypothesized that premature ventricular activation induces abnormal ventricular septal movement, LV dyssynchrony, and remodeling that may yield abnormal findings on echocardiography.[6,8] Some studies have also demonstrated finding of myocardial dyskinesia in segments that were precociously activated by accessory pathway in patients with WPW syndrome.[5,7,9]

Both of our patients had WPW displaying features of anteroseptal accessory pathway. Since it was asymptomatic and found at young age, first patient was followed up conservatively. Second case had palpitations and was referred for ablation therapy. In first patient, gradual disappearance of pre-excitation was observed, and systolic bulging of muscular septum into RV also decreased with significant improvement in LV systolic function. This finding gave rise to thought that pre-excitation may have been a cause of septal aneurysm, rather than result.

In patients with muscular septal aneurysm, bulging of interventricular septum into RV alters M-mode and 2-dimensional measurements, which may lead to misdiagnosis as DCM. In presently described patients, EF calculated according to Simpson's method was 40% and 46%, with CTR of 0.64 and 0.54.

DCM in patients with WPW syndrome is uncommon. Although it may occur in symptomatic WPW patients with sustained tachyarrhythmia, recent observations suggest that significant LV dysfunction may also arise in the absence of incessant tachyarrhythmia.[5–9] In reported series, almost all of the patients with non-

tachyarrhythmia-type DCM had paraseptal accessory pathway. It is thought that premature ventricular activation over these accessory pathways induces septal wall motion abnormalities and ventricular dyssynchrony.[5–9] Both of our patients had WPW with features of anteroseptal accessory pathway and presented as DCM. First patient had intermittent WPW on 24-hour Holter electrocardiogram. We observed resolution of LV dysfunction and pre-excitation in first case. Emmel et al.[5] reported 4 cases of DCM associated with WPW syndrome without sustained tachycardia. They observed resolution of cardiomyopathy with catheter ablation in 2 patients, and spontaneous disappearance of pre-excitation in 1. Park et al.[8] reported that LV dyssynchrony was different according to location of accessory pathway, and that dyssynchronous LV contraction improved immediately after catheter ablation in patients with pre-excitation syndrome and in canine models.

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Anahtar sözcükler: Kardiyomiyopati; dissenkroni; pre-eksitasyon; septal anevrizma.