Biventricular noncompaction and mitral cleft

Biventriküler "noncompaction" ve mitral kleft

Introduction

Noncompaction cardiomyopathy (NCC) is a rarely seen type of congenital cardiomyopathy. It is caused by the defective embryonic migration of myocardial fibers (1). It is frequently complicated with heart failure, arrhythmias and embolic events. In this report, we presented a case with biventricular noncompaction accompanying a cleft mitral valve.

Case Report

A 20-year-old male patient was admitted to our clinic with exertional dyspnea, fatigue and palpitations for the last 5 months. Arterial blood pressure was 120/60 mmHg, temperature was 36.6°C, and heart rate was 96 bpm and regular. A 3/6 grade of systolic murmur was heard over apical region. A normal sinus rhythm, left axis deviation, and signs of left ventricular hypertrophy were seen on ECG. The transthoracic echocardiography (TTE) (Vivid 3, GE Medical Systems) revealed double-contoured and hypertrabeculated ventricular myocardium involving both the ventricles. Deep trabeculations were present in noncompacted region. The ratio of noncompacted to compacted part was >2.0. Blood flow was detected inside deep trabeculations on color-Doppler echocardiography. Left ventricular apico-septal hypokinesia was also present. Left ventricular ejection fraction was 54%. The left atrium was larger than normal and moderate regurgitation flow through anterior mitral valve was seen. A cleft was suspected on anterior mitral leaflet in apical four-chamber and parasternal short-axis views, because the leaflet was seen as a two separate structure including a mitral regurgitation jet between two on color-Doppler echocardiography. Other valves were normal. For further anatomical investigation, transesophageal echocardiography (TEE) was performed (Vivid 3, GE Medical Systems). A cleft was present on anterior mitral leaflet with a moderate mitral regurgitation confirming the TTE findings (Fig. 1, Video 1, 2. See corresponding video/movie images at www.anakarder.com). No defect was seen in interatrial and interventricular septum. Deep trabeculations and blood flow in these deep trabeculations suggesting the noncompaction were observed in both ventricles (Video 3. See corresponding video/movie images at www.anakarder.com).

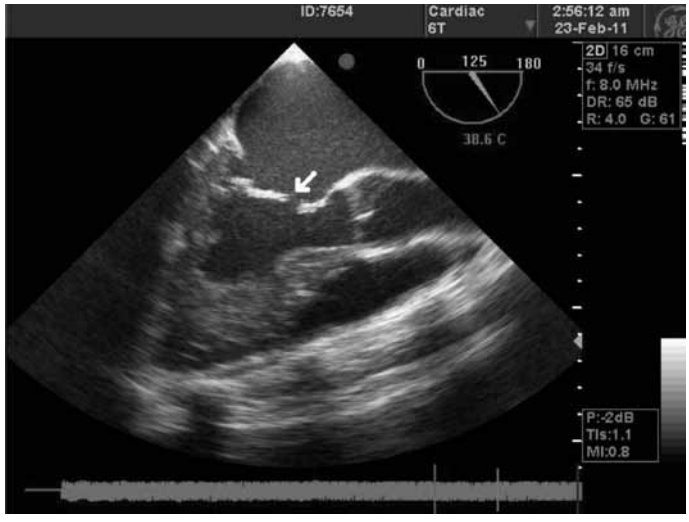

Figure 1. Transesophageal echocardiography image of anterior mitral leaflet cleft (white arrow)

Discussion

The NCC was defined by Chin and his colleagues in 1990 (1). This genetic cardiomyopathy is caused by a pause in interaction of myocardial fibers resulting in a disordered endomyocardial morphogenesis during embryonic survival (2). Echocardiographic diagnostic criteria are: after the exclusion of structural heart abnormalities; numerous, prominent trabeculations and deep intratrabecular recesses, intraventricular blood reaching recesses with color-Doppler, presence of two layers of compacted and non-compacted walls in parasternal short-axis, and non-compacted/compacted ratio > 2.0 (3). In the present case, all of these criteria were present. In some reports, magnetic resonance imaging is recommended. We didn't perform magnetic resonance imaging due to some technical problems and the fact that all criteria were present eliminating any suspicion.

The NCC can lead to serious clinical conditions such as heart failure, malignant arrhythmia and embolism (3, 4). Right ventricular involvement has been found in <50% of the cases, but the left ventricular involvement rate is 89%. It may also be biventricular (5).

Pediatric incidence of mitral cleft, a rarely seen valve anomaly, is 1:1340. Frequently the anterior leaflet of the mitral valve is involved. It is caused by incomplete expression of endocardial cushion (6). In our case, mitral cleft was also present. There are no established echocardiographic criteria for mitral cleft. In our case, the anterior mitral leaflet was constituted by two separate parts, resembling tricuspid mitral valve. Presence of moderate mitral regurgitation between these parts prompted us to define it as mitral cleft. The mitral cleft is frequently associated with other endocardial cushion defects, such as ostium primum atrial septal defect, atrioventricular septal defect (7). Until present, only one case with biventricular noncompaction and cleft mitral valve has been reported. There were also complete atrioventricular block and atrial septal aneurysm previously in that case, which was reported by Dağdeviren et al. (8). Besides, there is one other case of NCC, which was reported to be accompanying another valvular abnormality, Ebstein anomaly (9).

Conclusion

In a patient with cardiomyopathy with valvular abnormalities, the NCC should be kept in mind. The relation between NCC and valvular abnormalities needs to be delineated in further prospective studies.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Authors' contributions

ZI performed echocardiographic studies. ZI, SC, OY, MU analyzed and interpreted the clinical data, and ZI was a major contributor to writing the manuscript. All authors read and approved the final version of the manuscript.

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Video 1. Transesophageal echocardiography showing anterior mitral leaflet cleft

Video 2. Color-Doppler echocardiography showing mitral regurgitation

Video 3. Trabeculations in both ventricles

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Nadir bir birliktelik: Atriyoventriküler nodal yeniden giriş taşikardisi ve Mahaim aksesuvar yol aracılı atriyoventriküler taşikardi

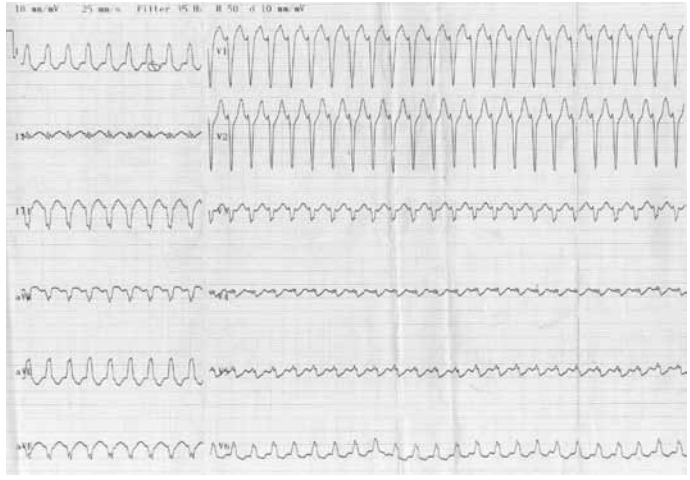
A rare coexistence: Atrioventricular nodal reentry tachycardia and Mahaim accessory pathway mediated atrioventricular tachycardia

Giriş

Mahaim yolları aksesuvar yolların yaklaşık %3'ünü oluşturur. Genel popülasyonda görülme sıklığı 0.5-1:10000'dir. Sol dal bloğu morfolojisinde atriyoventriküler yeniden giriş taşikardisinden (AVNRT) sorumludurlar (1, 2). Farklı aksesuvar yollar ve çift atriyoventriküler (AV) iletim ile birlikte olabilir, Ancak AV düğüm yeniden giriş taşikardi (AVNRT) ile birlikteliği oldukça nadirdir (3). Bohora ve ark.larının (4) yaptığı bir çalışmada elektrofizyolojik çalışma yapılan 510 hastanın 15'inde Mahaim tip aksesuvar yol saptanmış. Bu hastalardan 1 tanesinde çift AV düğüm fizyolojisi ve AVNRT tespit edilmiş (4). Bu vaka raporunda 24 yaşında kadın hastada Mahaim aksesuvar yolu taşikardisine eşlik eden atriyoventriküler düğüm yeniden giriş taşikardisinin de bulunduğu bir olgu sunulmuştur.

Olgu Sunumu

Beş yıldır çarpıntı yakınması olan 24 yaşındaki kadın hasta, son zamanlarda tıbbi tedaviye rağmen çarpıntılarının sıklığında artış olması üzerine başka bir merkezden herhangi bir belgelenmiş supraventriküler (SVT) ya da ventriküler taşikardi kaydı olmadan elektrofizyolojik çalışma (EFÇ) için hastanemize yönlendirilmiş. Hastanın fizik muayenesi ve laboratuvar bulguları normaldi. Standart 12 derivasyon elektrokardiyogramda (EKG) sinüs ritminde, normal PR aralığı (151 msn) ile birlikte normal QRS süresi (103 msn) mevcuttu. Ayrıca iki boyutlu transtorasik ekokardiyografide sol ventrikül çapları ve sistolik fonksiyonları ile kapak fonksiyonlarının normal olduğu izlendi. Hastamız klinikte izlenirken, gelişen çarpıntı yakınması ile çekilen 12 derivasyonlu EKG de sol dal bloğu morfolojisinde geniş QRS'li taşikardi saptandı (Şekil 1). Bilgilendirilmiş onamı alındıktan sonra hasta EFÇ'ye alındı. Standart elektrot kateterler yüksek sağ atriyuma, sağ ventriküler apeksine ve His hüzmesine yerleştirildi. Sinüs ritmindeki hastada, inkremental atriyal uyarı sırasında AH aralığı gittikçe kısaldığı ve sol dal bloğu morfolojisinde preeksitasyonun açığa çıktığı izlendi. Atriyal uyarı ile aynı zamanda sol dal bloğu morfolojisinde taşikardi indüklendi (Şekil 2). İntrakardiyak kayıtlarda taşikardi esnasında sağ dalın His hüzmesinden önce aktif olduğu izlendi. Aynı zamanda programlı atriyal uyarı ile VA aralığı 45 msn olan AVNRT ile uyumlu SVT indüklendi (Şekil 3). Sağ posteriyor yaklaşım ile AVNRT başarılı şekilde ablate edildi. Mahaim aksesuvar yol aracılı atriyoventriküler yeniden giriş taşikardisi için triküspit anülüsün sağ lateral bölgesinde, Mahaim aksesuvar yol potansiyelinin olduğu bölgeye, radyofrekans uygulandı ve başarılı ablasyon sağlandı. Radyofrekans uygulama sırasında aksesuvar yolun artmış otomatisitesine bağlı hızlanmış ritim izlendi. Atriyal uyarı ile tekrar preeksitasyon indüklenemedi. İşlem sonrası yüzey EKG'si normal



Şekil 1. Çarpıntı esnasında çekilen 12 derivasyonlu elektrokardiyografi: sol dal bloğu, geniş QRS ancak QRS süresi <0.15sn, DI'de R dalgası, V1'de rS kompleksi, prekordiyal geçişin V4 ve daha sonrasında olması, kalp hızının 130-170 arasında olması gibi Mahaim aksesuvar yolunun iştirak ettiği taşikardilere benzer özellikler izlenmektedir