Unusual association of multiple congenital left ventricular diverticulum and cerebrovascular events in an adult

Erişkinde ender görülen çok sayıda konjenital sol ventrikül divertikülü ve serebrovasküler olay birlikteliği

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Summary– Congenital ventricular diverticulum is a rare and usually asymptomatic cardiac malformation which can cause major complications such as systemic thromboembolism, infective endocarditis, cardiac rupture, heart failure, arrhythmia and sudden death. We present a case with multiple congenital left ventricular diverticulum admitted to hospital with sudden onset right-sided hemiplegia and dysarthria.

Congenital ventricular diverticulum is usually an Ooutward pouch formation of the ventricle, and is most commonly seen in the left ventricle. It is frequently accompanied by other cardiac abnormalities, and is most commonly diagnosed during early childhood. No accompanying congenital malformations are seen in approximately 30% of cases.^[1] Due to its asymptomatic course, diagnosis of left ventricular diverticulum is difficult. Therefore, it is usually coincidentally detected during routine transthoracic echocardiographic evaluation or cardiac catheterization.

In this report, we present a case with multiple congenital left ventricular diverticula admitted to hospital with sudden onset stroke in the absence of midline defects or congenital heart disease.

CASE REPORT

A 33-year-old female patient was admitted to our clinic for investigation of the etiology of an ischaemic cerebrovascular event. Her medical history was unremarkable, with no orthostatic hypotension, seizure

Özet– Doğumsal sol ventrikül divertikülü nadir görülen bir kardiyak anomali olup genellikle asemptomatik iken, sistemik tromboemboli, enfektif endokardit, kardiyak rüptür, kalp yetersizliği, aritmi ve ani kardiyak ölüm gibi ciddi komplikasyonlara neden olabilir. Bu yazıda, ani başlangıçlı sağ hemipleji ve dizartri şikayeti ile başvuran çoklu sol ventrikül divertiküllü saptanan bir olgu sunuldu.

disorders or diabetes mellitus. She denied prior use of neuroleptics or oral contraceptives.

Abbreviation:

She had no family history of heart disease or neurological disorders. On physical examination, her blood pressure was 120/70 mmHg and heart rate 86 bpm. Cardiac examination was normal. Blood tests including glucose, glycosylated haemoglobin A1c, thyroid and parathyroid function test, rheumatoid factor, lupus anticoagulant, antiphospholipid antibody, antinuclear antibody, antineutrophil cytoplasmic antibody, anti-Ro/La antibody, and peripheral blood morphology were either normal or unremarkable. The 12-lead electrocardiogram (ECG) showed sinus rhythm of 86 bpm, T wave abnormality in leads V3-4 and incomplete left bundle branch block. A 24-hour ECG recording was performed and it did not reveal any signs of arrhythmia. Carotid artery Doppler ultrasonography and transesophageal echocardiography were performed to rule out the possibility of carotid artery disease, patent foramen ovale or atrial septal defect, and the results were normal. Transthoracic echocardiog-

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ECG Electrocardiogram

raphy revealed prominent trabecular meshwork and deep trabecular sinusoids in the posterolateral wall, consistent with diverticulum (Figure 1). Coronary angiography, which was performed to rule out possible coronary artery disease, showed normal epicardial arteries. Left ventriculography revealed multiple contractile diverticula, originating from the lateral wall and left ventricular apex (Figure 2). Cardiac magnetic resonance imaging (MRI) was performed and diverticula of dimensions 7.0x2.5 cm and 1.2x1.0 cm were detected on the lateral wall of the left ventricle at the

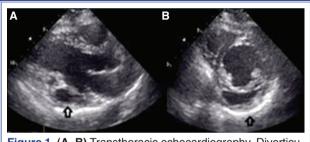


Figure 1. (A, B) Transthoracic echocardiography. Diverticulum within the posterolateral wall (black arrows) Parasternal long axis and short axis.



Figure 2. Left ventriculography. Multiple diverticula, orginating from the lateral wall and apex of the left ventricule (black arrows). (A) Diastole (B) Systole.

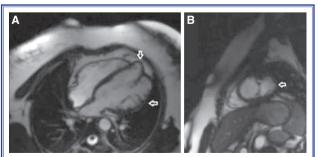


Figure 3. Cardiac magnetic resonance imaging. Multiple diverticula, orginating from the posterolateral wall and apex of the left ventricule. (A) Apical 4 chamber views (B) mid-basal short axis views.

apex (Figure 3). Upon diagnosis of multiple congenital left ventricular diverticula, the case was presented to the cardiology and cardiovascular surgery council. Pharmacotherapy with warfarin, metoprolol and ramipril was initiated. Outpatient follow-up was recommended, and at the end of one-year follow-up, no cardiac symptoms or systemic embolism were discovered.

DISCUSSION

Left ventricular diverticulum is a rare cardiac abnormality. The incidence of left ventricular diverticulum is reported to be 0.04% in the general population, and approximately 0.02% in a previously reported consecutive pediatric autopsy series.^[2,3] Ventricular diverticula are most frequently classified as muscular and fibrous. Although their etiology is not clear, an embryological developmental defect has been proposed.^[4,5] Muscular diverticula usually bud from the left ventricular apex with a narrow stalk. They are frequently associated with other cardiac or extracardiac congenital abnormalities. Fibrous diverticula originate from the apex or base of the ventricle near the atrioventricular valvular ring.^[6] Left ventricular diverticulum needs to be differentiated from a ventricular aneurysm or pseudoaneurysm. A narrow neck and synchronous contractility along with all three ventricular layers indicate a diverticulum.^[7] In contrast, an aneurysm or pseudo aneurysm shows akinesia or paradoxical contractility of the outpouching with a broad neck myocardium in an aneurysm and a narrow neck/pericardium in a pseudo aneurysm. They are not associated with cardiac or extracardiac congenital defects. Patients with cardiac diverticulum are usually asymptomatic. However, there are reports which highlight its association with arrhythmia, chest pain, embolic events, heart failure, and sudden death due to diverticulum rupture.^[8] In addition, as seen in our case, they can cause systemic embolisms. Therefore, the management of these cases is not clearly defined, and treatment of this abnormality in asymptomatic patients is controversial. There have been a range of treatments, such as antiplatelets, anticoagulants, and even surgical removal. Diverticular resection is often indicated for patients who have concomitant congenital heart disease or other malformations. Anticoagulant therapy is indicated for systemic embolisms.^[9] The prognosis depends on the severity and the degree of concomitant malformations. Isolated and asymptomatic cases usually have a benign course.[10]

The present case highlights the importance of detailed and careful transthoracic echocardiography in evaluation of patients who present to a cardiology clinic with systemic embolism, in which left ventricular diverticulum can be a rare, but significant cause.

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Anahtar sözcükler: Aort hastalıkları/komplikasyon; serebrovasküler olay/etiyoloji; divertikül; transözofajiyal ekokardiyografi; emboli.