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Isolated Cardiac Sarcoidosis in a Patient with a Rare Coronary Anomaly Presenting with MINOCA and Heart Block: The Devil Is in the Detail

MINOCA ve Kalp Bloğu ile Gelen Nadir Bir Koroner Anomalili Hastada İzole Kardiyak Sarkoidoz: Şeytan Ayrıntıda Gizli

CASE REPORT
OLGU SUNUMU

ABSTRACT

A middle-aged pre-menopausal female presented with shortness of breath and syncope. She had a past history of acute onset chest pain with elevated cardiac enzyme, regional wall motion abnormality on echocardiography, and a coronary anomaly in angiogram. She was being treated as a case of coronary artery disease. On current evaluation, she had right bundle branch block with intermittent 2 : 1 AV block on ECG and a hyperechoic and hypokinetic interventricular septum with moderate left ventricular systolic dysfunction on echocardiography. Coronary angiogram revealed hyperdominant left anterior descending with right coronary artery ostial atresia. The patient was diagnosed to have cardiac sarcoidosis on the basis of epicardial late gadolinium enhancement (LGE) on MRI and increased use of 68-Gallium DOTANOC PET scan. Patient underwent dual-chamber ICD implantation and then steroids were started.

Keywords: Cardiac sarcoidosis, cardiomyopathy, inflammation, left ventricular dysfunction

ÖZET

Orta yaşlı, premenopozal bir kadın hasta, nefes darlığı ve senkop ile başvurdu. Geçmişte kardiyak enzim yüksekliği ile akut başlangıçlı göğüs ağrısı öyküsü, ekokardiyografide bölgesel duvar hareket anormalliği ve anjiyogramda koroner anomalisi vardı. Koroner arter hastalığı vakası olarak tedavi ediliyordu. Mevcut değerlendirmede, EKG'de aralıklı 2:1 AV bloklu sağ dal bloğu (RBBB) ve ekokardiyografide orta derecede LV sistolik disfonksiyonu olan hiperekoik ve hipokinetik interventriküler septum vardı. Koroner anjiyogramda, RCA ostial atrezili hiperdominant LAD görüldü. Hastaya MRI'da epikardiyal geç gadolinyum kontrastlanması (LGE) ve 68-Galyum DOTANOC PET taramasında artan tutuluma dayalı olarak kardiyak sarkoidoz tanısı kondu. Hastaya çift odacıklı ICD implantasyonu yapıldı ve ardından steroid başlandı.

Anahtar Kelimeler: Kardiyak sarkoidoz, kardiyomiyopati, enflamasyon, sol ventrikül disfonksiyonu

Patients presenting with MINOCA (Myocardial Infarction with No Obstructive Coronary Arteries) should be evaluated meticulously to arrive at a timely diagnosis of potentially reversible conditions, like cardiac sarcoidosis in our case, so that when treated with steroids at an early stage, irreversible changes may be prevented. Cardiac sarcoidosis usually presents with heart block, ventricular tachycardia, and heart failure. Cardiac sarcoidosis manifesting as MINOCA is quite rare. Anomalous origin of coronary arteries is also a rare phenomenon that is otherwise asymptomatic and it mostly comes to light while performing coronary angiograms. We report a patient with a past history suggestive of acute coronary syndrome, who currently presented with shortness of breath and syncope. She was diagnosed to have a hyperdominant left anterior descending (LAD) artery with atresia of right coronary artery (RCA) ostium, and on evaluation turned out to have cardiac sarcoidosis as well. We report this case in view of the rarity of two things: a very rare coronary anomaly and a rare presentation of cardiac sarcoidosis with MINOCA.

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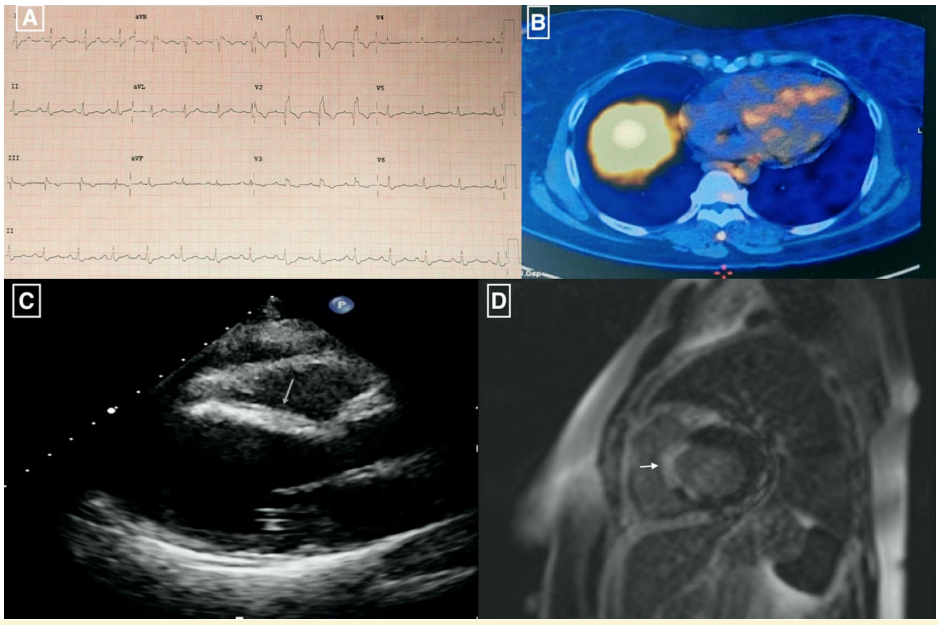

Figure 1. A-D. (A) Electrocardiogram showing RBBB and deep Q in leads V1 and V2. (B) 68-Ga DOTANOC PET scan showing increased uptake in the IVS. (C) Transthoracic echocardiography in parasternal long-axis view showing hyperechogenic IVS. (D) T1 scout image of cardiac MRI showing LGE in septum. RBBB, right bundle branch block; 68-Ga, 68 Gallium; LGE, late gadolinium enhancement.

Case Report

A 48-year-old female, hypertensive and hypothyroid, with a history of an episode of acute onset retrosternal chest pain 3 years back, was evaluated elsewhere and diagnosed as acute coronary syndrome based on elevated troponin I levels and wall motion abnormality on echocardiography. Coronary angiogram was performed, which revealed a coronary anomaly with no significant obstructive disease. CT coronary angiogram did not reveal any plaques, and coronary artery calcium score was zero. No further evaluation was done and she was managed as a case of coronary artery disease. She was put on dual antiplatelet drugs (aspirin and clopidogrel), ACE inhibitor, and beta-blocker. She remained asymptomatic for the next two-and-a-half years. She presented to our center with dyspnea on exertion of NYHA (New York Heart Association) class II for 6 months and 2 episodes of syncope. On examination, her vitals were stable, and cardiovascular system examination was essentially unremarkable. Her electrocardiogram revealed right bundle branch block (RBBB), Q waves in leads V1 and V2, with a PR interval of 190 ms (Figure 1). Twenty-four hours ECG monitoring revealed intermittent 2:1 AV block. Echocardiography demonstrated moderate left ventricular (LV) systolic dysfunction, hyperechoic interventricular septum, and hypokinesia of the entire interventricular septum, with an ejection fraction of 40% (Figure 1; Videos 1-3*). The chest x-ray was normal. Her blood counts, biochemistry (except C-reactive protein), and lipid profile were within normal limits.

The patient underwent coronary angiogram which showed hyperdominant LAD artery continuing as posterior descending artery (PAD). The ostium of the RCA was atretic with hypoplastic proximal RCA supplied by a branch from mid-LAD artery. Postero-lateral ventricular branch arose from the left circumflex artery (Figure 2). There was no obstructive coronary artery disease. Electrophysiological study revealed prolonged HV interval (96 ms) and infra-Hisian block on atrial pacing. Cardiac MRI revealed areas of focal thickening and thinning of the interventricular septum along with LGE in the interventricular septum, LV apex, and right ventricular lateral wall. The LV ejection fraction in cardiac MRI was 34%. Based on these MRI findings and infra-Hisian conduction disease, a provisional diagnosis of cardiac sarcoidosis was made. Subsequently, 68-Ga (68 Gallium) DOTANOC PET CT scan showed increased uptake in interventricular septum suggestive of acute inflammatory pathology (Figure 1). Tc99 sestamibi myocardial perfusion imaging revealed severely hypoperfused basal and anterior septum. The CRP was elevated (8.3 mg/dL). Troponin I, ANA, ANCA, and ACE levels were normal. Mantoux test revealed anergy and viral markers were negative. Contrast-enhanced chest CT did not show any significant mediastinal lymphadenopathy or lung parenchymal disease. Endomyocardial biopsy was non-diagnostic and showed focal replacement fibrosis. There were no features to suggest extracardiac involvement of sarcoidosis. Differential diagnosis for such presentation included giant cell myocarditis, hypersensitivity myocarditis, and lymphocytic myocarditis. Giant cell myocarditis usually has a fulminant course, hence unlikely to the etiology in our case. Hypersensitivity myocarditis is associated with drug intake and eosinophilia, which was not present in our case.

ABBREVIATIONS

RBBB	Right bundle branch block
LGE	Late gadolinium enhancement
MINOCA	Myocardial Infarction with No Obstructive Coronary Arteries
LAD	Left anterior descending
RCA	Right coronary artery
NYHA	New York Heart Association
PDA	Posterior descending artery

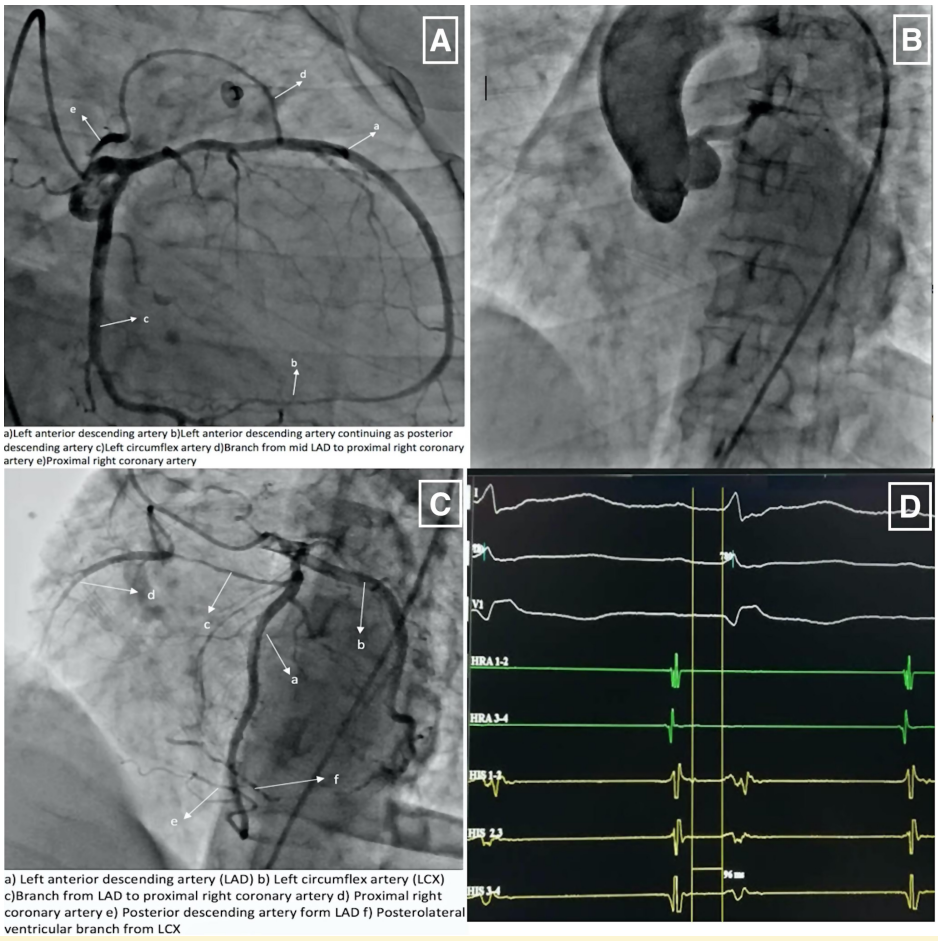

Figure 2. A-D. (A) Left coronary angiogram in Right anterior oblique (RAO) caudal view. (B) Aortic root angiogram in Left anterior oblique (LAO) cranial view. (C) Left coronary angiogram in LAO cranial view. (D) Electrophysiology study showing prolonged HV interval.

Lymphocytic myocarditis may be either because of viral or autoimmune etiology. Patient neither had any viral prodrome prior to initial presentation nor any features suggestive of autoimmune disease. Myocardial biopsy lacked any inflammatory cells and DOTONAC scan which has a high affinity for sarcoid granulomas was positive. Based on these findings, a diagnosis of cardiac sarcoidosis was made.

Since the patient had an indication for both pacing and primary prevention of sudden cardiac death (in virtue of LV ejection fraction of 34% in cardiac MRI and LGE on MRI), she underwent dual-chamber implantable cardiac defibrillator implantation. Oral steroids were started since there was evidence of ongoing myocardial inflammation. Optimal medical therapy was initiated for heart failure with metoprolol succinate, ramipril, and spironolactone. The patient had an uneventful post-procedural hospital stay. She did not have any further episodes of syncope and showed significant improvement in dyspnea.

Discussion

Anomalies of coronary arteries are rare with an incidence of 0.2-1.3% in angiographic series and 0.3% in autopsy series.[1] Hyperdominant LAD artery, also known as anomalous origin of PDA from LAD, in which LAD coronary artery continues as PDA, is a rare anomaly with 19 cases reported to date.[2] Wrap around LAD artery is one where the vessel extends beyond the apex to supply the apical inferior wall, whereas hyper-dominant LAD extends further to give rise to PDA. Our patient had hyperdominant LAD artery with atresia of ostio-proximal portion of RCA. Hypoplastic proximal RCA was supplied by a branch from mid-LAD artery. Postero-lateral ventricular branch arose from an obtuse marginal artery. To the best of our knowledge, there is only a single report of a similar combination of anomalies reported previously.[3] Occlusion of hyperdominant LAD artery would be catastrophic as blood supply to a large myocardial territory will be jeopardized.

In our case, the presentation of the patient with acute chest pain with RBBB, regional wall motion abnormality, and the presence of a coronary anomaly deceived the actual diagnosis and hindered further evaluation. The patient had MINOCA during the initial presentation based on diagnostic criteria laid down by ESC working group position paper on MINOCA.[4] The posterobasal segment of interventricular septum is supplied by PDA and the rest of interventricular septum is by LAD artery. On the careful assessment of regional wall motion abnormality, the entire interventricular septum was involved, which did not conform to any particular coronary artery territory. Even though acute coronary event involving a hyperdominant LAD may lead to hypokinesia of the entire septum, this is unlikely in this case as

there was no obstructive coronary artery disease and coronary artery calcium score was normal. Also, interventricular septum was hyperechogenic. 68-Ga DOTANOC PET is very useful in diagnosing cardiac sarcoidosis, as type 2 somatostatin receptors are overexpressed in sarcoid granulomas, while it is not found in normal myocardium. Ga-68 DOTANOC PET scan has a sensitivity and specificity of 100% and 88%, respectively.[5,6] Based on the combination of hyperechogenic, hypokinetic septum with ventricular dysfunction, 2:1 AV block, subepicardial LGE on MRI, increased septal uptake in 68-Ga DOTANOC PET, and a negative mantoux test, a diagnosis of cardiac sarcoidosis was made. Hence the initial presentation of the patient with MINOCA was due to the invasion of sarcoid granuloma in the ventricular septum, which was misdiagnosed and treated as coronary artery disease. The presence of a rare coronary anomaly further compounded the clinical picture and diverted attention from the exact etiology of MINOCA.

Cardiac sarcoidosis usually presents with AV blocks, heart failure, and arrhythmias. In a study of 42 consecutive patients with cardiac sarcoidosis, 7% of the patients had angina pectoris.[7] However, the presentation of cardiac sarcoidosis with acute chest pain is quite rare. Coronary involvement may be in the form of coronary sarcoidosis leading to epicardial coronary artery stenosis, coronary aneurysm, spontaneous coronary artery dissection, or microvascular involvement due to myocardial sarcoid granuloma.[8-11] There is a previous report of a patient with pulmonary sarcoidosis on treatment, presenting with non-ST elevation myocardial infarction and normal epicardial coronary arteries.[9] But, MINOCA as the first manifestation of sarcoidosis, followed by a long asymptomatic period, and finally resurfacing with heart failure symptoms and conduction abnormalities to our knowledge has never been reported. Since the patient had an indication of permanent pacemaker and ICD implantation based on the Heart Rhythm Society consensus statement of diagnosis and management of arrhythmias in cardiac sarcoidosis, dual-chamber ICD was implanted.[12] Since the patient had evidence of active disease, an oral steroid was initiated.

Conclusion

Meticulous assessment of regional wall motion abnormality is of paramount importance in the evaluation of cases presenting with acute chest pain. Young patients presenting with AV blocks and unexplained ventricular dysfunction should be evaluated for sarcoidosis. Hyperdominant left coronary artery with RCA ostial atresia is a rare benign coronary anomaly. Inflammatory cardiomyopathy like cardiac sarcoidosis should be considered in the differential diagnosis of MINOCA in premenopausal females without conventional coronary artery disease risk factors.

*Supplementary video files associated with this article can be found in the online version of the journal.

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