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Mesenteric Artery Embolism During Coronavirus Disease-19 (COVID-19) İn A Patient Previously Diagnosed With Fabry: A Case Report

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ABSTRACT

We report a unique case of superior mesenteric artery embolism that developed in a patient with COVID-19 who was followed up with the diagnosis of Fabry Disease. On the 12th day of COVID-19 illness, atrial fibrillation and superior mesenteric artery embolism were detected in a 56-year-old male patient who was previously diagnosed with Fabry Disease. We did not find a similar patient in the literature. In the coexistence of Fabry Disease and COVID-19, risk of arrhythmia such as atrial fibrillation, which is the most common arrhythmia in both diseases, increases further. COVID-19 can cause arterial and venous thrombosis. In the coexistence of Fabry Disease and COVID-19, care should be taken against increased thrombogenicity, arrhythmia (such as atrial fibrillation) and thromboembolism that may develop due to them.

Keywords: Fabry disease, COVID-19, Atrial fibrilation, Superior mesenteric artery, Embolism

Introduction

Fabry disease (FD) is an X-linked lysosomal storage disease caused by mutations in the gene encoding the enzyme α - galactosidase A (α - Gal A) (1). This defect leads progressive enzymatic to the accumulation of glycosphingolipids, mainly globotriaosylceramide (Gb3) globotriaosylsphingosine (lyso-Gb3), in lysosomes and causes multisystemic problems, including neurological, ocular, skin, brain, renal and cardiac symptoms (2). As a result of cardiac involvement in FD; left ventricular hypertrophy (LVH - the most prominent finding) / right ventricular hypertrophy (RVH), cardiomyopathy, aortic and mitral valve abnormalities, arrhythmias, coronary artery disease, heart failure, aortic root dilatation may develop (3). Arrhythmia (including atrial fibrillation (AF) and ventricular tachycardia) may occur in 27-42% of men and in 27% of female patients with FD (4). Coronavirus Disease-19 (COVID-19), caused by severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2), was described as a global pandemic by the World Health Organization in March 2020. Patients with a history of cardiovascular disease are at higher risk for COVID-19 complications (5).

COVID-19 can cause arterial and venous thrombosis (6). Cardiac arrhythmias can occur during or after COVID-19. Atrial arrhythmias such as AF are the most frequently reported arrhythmias in COVID-19 patients (7).

Case Report

On the 12th day of COVID-19 illness, a 56-year-old male patient who was previously diagnosed with FD started the complaint of palpitations and then abdominal pain. The patient, who was followed up with symptomatic treatment at home, was diagnosed with COVID-19 by oropharyngeal swab and was not receiving anticoagulant therapy. Superior mesenteric artery (SMA) embolism was detected in the contrastenhanced abdominal computed tomography taken with the preliminary diagnosis of acute mesenteric ischemia in the patient who applied to the emergency department on the 2nd day with the complaint of abdominal pain (Figure 1). AF was detected in the electrocardiography (ECG) of the patient, who had no previous palpitations (Figure 2A). Left ventricular concentric hypertrophy was observed echocardiography and left atrial diameters were

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Fig. 1. Occlusion of the superior mesenteric artery (SMA)

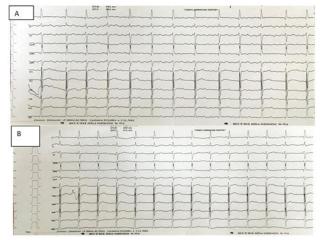


Fig. 2. 2A:Atrial fibrillation (AF) detected in the first electrocardiography (ECG), 2B: ECG control after cardioversion

normal (Figure 3). The patient, whose respiratory system examination did not reveal any significant pathology, was taken to emergency operation because of severe abdominal pain and acute mesenteric ischemia, and necrotic bowel resection was performed. Transesophageal echocardiography was performed on the patient whose postoperative AF continued. Amiodarone infusion was started for medical cardioversion in the patient who did not have intracardiac thrombus, and sinus rhythm could not be achieved. SR was achieved with cardioversion (Figure 2B). Medical follow-up and treatment decision was made for the lesions detected in the SMA angiography performed in the patient who had occasional abdominal pain (Figure 4). Antiarrhythmic and anticoagulant treatment of the patient whose complaints decreased, was arranged in accordance with the guidelines and he was discharged. No new thrombotic events were observed in the patient who continued at the 1st, 3rd and 6th month controls. AF was not observed in the 24-hour Holter rhythm.

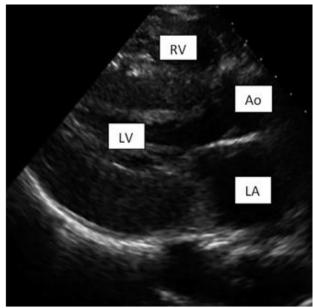


Fig. 3. Echocardiographic images of the patient; left ventricular hypertrophy (LVH)



Fig. 4. Control mesenteric artery angiography and the lesion to the superior mesenteric artery

Discussion

Cardiac involvement is a common cause of morbidity and mortality in FD. Cardiomyopathy, myocardial infarction, hypertension and rhythm abnormalities may be seen (8). The most common ECG finding is the voltage criterion for LVH. Left ventricular hypertrophy is associated with arrhythmias therefore the degree of hypertrophy may be a predictor of arrhythmic events in Fabry cardiomyopathy (9). Atrial arrhythmias, e.g. AF is more common than ventricular arrhythmias. In a case series with more than 10 years of follow-up, 13% of patients had

paroxysmal AF; Predictors for AF were age, left atrial size, LV wall thickness, LV mass index, and angina (10). Our patient was male and he had significant LV hypertrophy.

Specifically, Gb3/lyso-Gb3 deposition in endothelial cell is a hallmark of FD, and endothelial dysfunction and thrombophilia have been suggested as mechanisms underlying vascular involvement in patients with classical FD (11). In the screening of 129 patients followed up with the diagnosis of FD in Italy, no patient was diagnosed with COVID-19 or reported complaints (12). Multiple venous and arterial thrombosis have been reported in COVID-19. Acute cerebrovascular disease was seen in 5.7% of 241 patients, and ischemia in the lower extremities and two fingers was observed in 1 patient (13). Cardiac complications of COVID-19 include myocarditis, arrhythmias, heart failure, and acute coronary syndrome (plaque rupture, coronary thrombosis, etc.) (14). Cardiac arrhythmias may occur during and after COVID-19. Atrial arrhythmias are the most commonly reported arrhythmias in COVID-19 patients (7). AF is typically treated with rate and rhythm control and with anticoagulation in patients who meet criteria and do not have contraindications due to the risk of bleeding (15). The rhythm control strategy can be achieved with synchronized cardioversion or antiarrhythmic drugs such as amiodarone (16).

In our patient, AF was converted to SR with electrical cardioversion and antiarrhythmic and anticoagulant therapy was administered in accordance with the guidelines.

We report a unique case of SMA embolism that developed in a patient with COVID-19 who was followed up with the diagnosis of Fabry Disease. We did not find a similar patient in the literature. In addition to LVH and other mechanisms that can be seen in patients with FD, the increased risk of arrhythmia in COVID-19 disease may also contribute to the formation of AF. In the coexistence of FD and COVID-19, care should be taken against increased thrombogenicity, arrhythmia (such as AF) and thromboembolism that may develop due to them. We think that anticoagulant therapy should be kept in mind in the management of COVID-19 in patients with thrombogenic diseases such as FD.

Authors Contribution: All authors contributed to the description and discussion of the case.

Protection of Humans and Animals: The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the 2013 Helsinki Declaration of the World Medical Association.

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References

- Desnick RJ, Brady R, Barranger J, Collins AJ, Germain DP, Goldman M, Grabowski G, Packman S, Wilcox WR. Fabry disease, an under-recognized multisystemic disorder: expert recommendations for diagnosis, management, and enzyme replacement therapy. Ann Intern Med. 2003 Feb 18;138(4):338-46.
- 2. Kampmann C, Baehner F, Whybra C, Martin C, Wiethoff CM, Ries M, Gal A, Beck M. Cardiac manifestations of Anderson-Fabry disease in heterozygous females. J Am Coll Cardiol. 2002 Nov 6;40(9):1668-74.
- 3. Kampmann C, Baehner F, Ries M, Beck M. Cardiac involvement in Anderson-Fabry disease. J Am Soc Nephrol. 2002 Jun;13 Suppl 2:S147-149.
- 4. Yousef Z, Elliott PM, Cecchi F, Escoubet B, Linhart A, Monserrat L, Namdar M, Weidemann F. Left ventricular hypertrophy in Fabry disease: a practical approach to diagnosis. Eur Heart J. 2013 Mar;34(11):802-8.
- 5. Madjid M, Safavi-Naeini P, Solomon SD, Vardeny O. Potential Effects of Coronaviruses on the Cardiovascular System: A Review. JAMA Cardiol. 2020 Jul 1;5(7):831-840.
- 6. Jose RJ, Manuel A. COVID-19 cytokine storm: the interplay between inflammation and coagulation. Lancet Respir Med. 2020 Jun;8(6):e46-e47.
- 7. Berman JP, Abrams MP, Kushnir A, Rubin GA, Ehlert F, Biviano A, et al. Cardiac electrophysiology consultative experience at the epicenter of the COVID-19 pandemic in the United States. Indian Pacing Electrophysiol J. 2020 Nov-Dec;20(6):250-256.
- 8. Acharya D, Robertson P, Kay GN, Jackson L, Warnock DG, Plumb VJ, et al. Arrhythmias in Fabry cardiomyopathy. Clin Cardiol. 2012 Dec;35(12):738-40.
- 9. Cox GF. Diagnostic Approaches to Pediatric Cardiomyopathy of Metabolic Genetic

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- Etiologies and Their Relation to Therapy. Prog Pediatr Cardiol. 2007;24(1):15-25.
- Shah JS, Hughes DA, Sachdev B, Tome M, Ward D, Lee P, et al. Prevalence and clinical significance of cardiac arrhythmia in Anderson-Fabry disease. Am J Cardiol. 2005 Sep 15;96(6):842-6.
- 11. Demuth K, Germain DP. Endothelial markers and homocysteine in patients with classic Fabry disease. Acta Paediatr Suppl. 2002;91(439):57-61.
- 12. Riccio E, Pieroni M, Limoneglli G, Pisani A. Impact of COVID-19 pandemic on patients with Fabry disease: An Italian experience. Mol Genet Metab. 2020 Sep-Oct;131(1-2):124-125.
- 13. Zhang Y, Xiao M, Zhang S, Xia P, Cao W, Jiang W, et al. Coagulopathy and antiphospholipid antibodies in patients with COVID-19. N Engl J Med. 2020;382(17):e38.

- 14. Babapoor-Farrokhran S, Gill D, Walker J, Rasekhi RT, Bozorgnia B, Amanullah A. Myocardial injury and COVID-19: possible mechanisms. Life Sci. 2020;253:117723.
- 15. January CT, Wann LS, Alpert JS, Calkins H, Cigarroa JE, Cleveland JC Jr, et al. 2014 AHA/ACC/HRS guideline for the management of patients with atrial fibrillation: a report of the American College of Cardiology/American Heart Association Task Force on practice guidelines and the Heart Rhythm Society. Circulation. 2014;130(23):e199–267.
- 16. Rattanawong P, Shen W, El Masry H, Sorajja D, Srivathsan K, Valverde A, et al. Guidance on Short-Term Management of Atrial Fibrillation in Coronavirus Disease 2019. J Am Heart Assoc. 2020 Jul 21;9(14):e017529.