LETTER TO THE EDITOR

DOI: 10.4274/nkmj.galenos.2024.69772
Nam Kem Med J 2024;12(2):142-144

Hemochromatosis as a Preventable Cause of Heart Failure: A Rare Case

Kalp Yetmezliğinin Önlenebilir Bir Nedeni Olarak Hemokromatozis: Nadir Bir Olgu

Mustafa EBİK[1], Efe YILMAZ[1], Muhammet GÜRDOĞAN[1], Fethi Emre USTABAŞIOĞLU[2], Yekta GÜRLERTOP[1]

[1]*Trakya University Faculty of Medicine, Department of Cardiology, Edirne, Turkey*
[2]*Trakya University Faculty of Medicine, Department of Radiology, Edirne, Turkey*

Keywords: Hemochromatosis, thalassemia major, heart failure, ventricular tachycardia, global longitudinal strain, cardiac magnetic resonance

Anahtar Kelimeler: Hemokromatozis, talasemi majör, kalp yetmezliği, ventriküler taşikardi, global longitudinal strain, kardiyak manyetik rezonans

To the Editor,

A 29-year-old male patient presented to the emergency department with palpitations and fainting sensations. Ventricular tachycardia was detected on electrocardiography and sinus rhythm was achieved with cardioversion. The patient's medical history revealed a diagnosis of thalassemia major (TM) at the age of 1 year, splenectomy at the age of 5 years, and blood transfusions at external centers. Upon examination of the patient's hospitalization records at our hematology clinic, it was reported that the T2* value in the interventricular septum was 20.3 msec in the cardiac magnetic resonance (CMR) examination with T2* sequence performed for iron accumulation on October 6, 2015, and clinical follow-up was recommended. During the 8-year period after this date, the patient underwent T2* sequence CMR examinations to assess myocardial iron loading. The results showed the values of 6 msec and 4.5 msec, respectively. Despite recommendations from the hematology clinic, the patient did not attend cardiology check-ups and did not continue the deferiprone 500 mg 3x2 treatment prescribed for iron chelation. Furthermore, the patient has not visited our hospital for the past 1.5 years. Echocardiography was performed on the patient who was admitted to the coronary intensive care unit. The results showed an ejection fraction of 40%, left ventricular (LV) dilatation, and global hypokinesia. Strain echocardiography was also performed on recorded images, which revealed a decrease in the LV global longitudinal strain value compared to normal, with a value of -17.7%. The interventricular septum revealed a more pronounced decrease in the strain value (Figure 1B). Coronary angiography was performed to determine the cause of VT. The coronary arteries were found to be normal (Figure 1C, 1D). The patient's serum ferritin level was greater than 2000 µg/L. In the cardiology council, with the participation of a hematology specialist, the patient's current and previous CMR findings were evaluated. The clinical condition was determined to be due to cardiomyopathy/heart failure caused by secondary hemochromatosis. Another CMR imaging was performed to investigate the cardiac iron load. CMR imaging revealed LV enlargement and decreased systolic function on 4-chamber cine images. The mean T2* value in the interventricular septum was less than 5 milliseconds (Figure 2). Based on these results, the patient was diagnosed with heart failure due to secondary hemochromatosis caused by cardiac iron accumulation resulting from frequent transfusions due to thalassemia. The patient, who received follow-up and treatment in the cardiology clinic, was prescribed ramipril 2.5 mg once daily, spironolactone 25 mg once daily, carvedilol 6.25 mg twice daily, and furosemide 40 mg twice weekly. Additionally, amiodarone 200 mg was

Address for Correspondence: Mustafa EBİK MD, Trakya University Faculty of Medicine, Department of Cardiology, Edirne, Turkey
Phone: +90 542 251 34 50 **E-mail:** mustafaebik@gmail.com **ORCID ID:** orcid.org/0000-0002-0585-0958
Received: 19.06.2023 **Accepted**: 10.02.2024

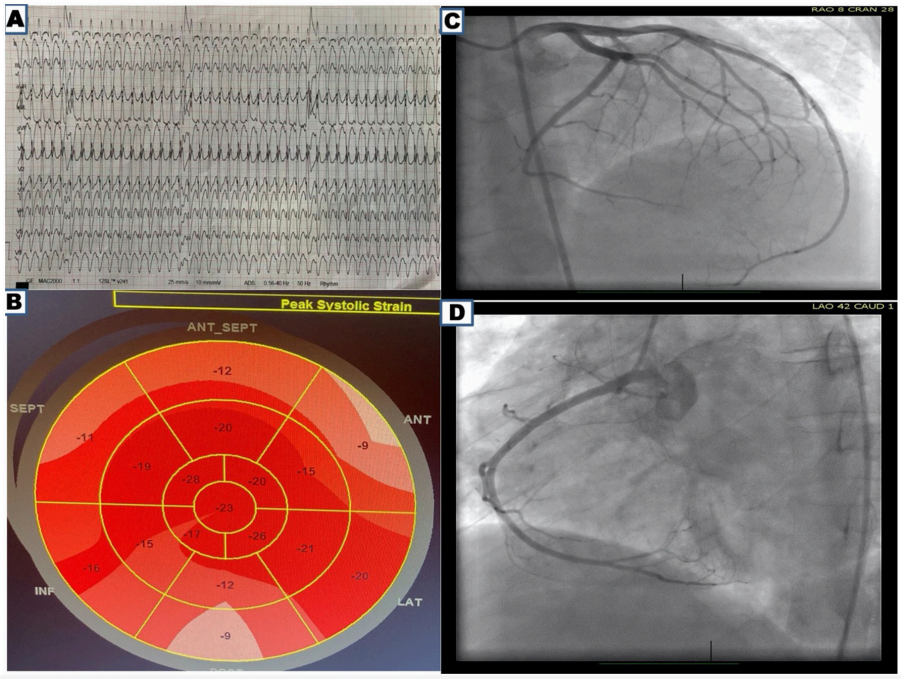

Figure 1. A) Ventricular tachycardia on ECG. B) Strain echocardiography shows a decrease in strain values, especially in the interventricular septum. C) Left system is normal in coronary angiography. D) Right system is normal in coronary angiography

ECG: Electrocardiography

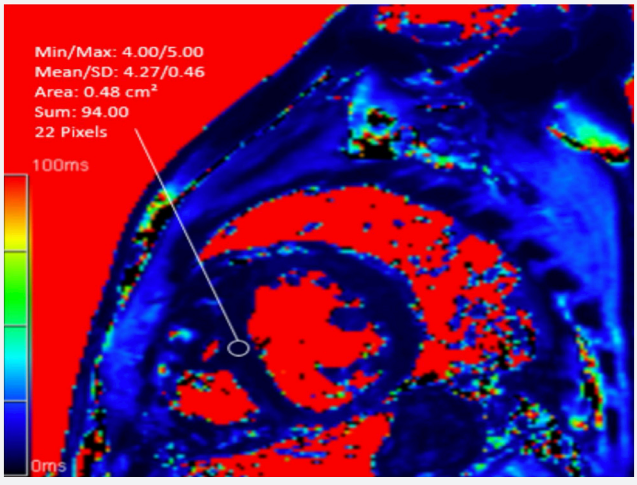

Figure 2. T2-star (T2*) mapping to show iron deposition in CMR imaging

CMR: Cardiac magnetic resonance

prescribed twice daily to prevent sudden cardiac death. The council performed cardioverter-defibrillator implantation for secondary prevention. Following a hematology consultation, iron chelation therapy was initiated with deferiprone 500 mg taken three times daily. The patient was discharged with a recommendation for outpatient clinic follow-up.

This case highlights the potential development of cardiac dysfunction due to myocardial iron loading in patients with frequent blood transfusions, such as those with TM. Therefore, it is important for patients to undergo periodic checks for cardiac hemochromatosis, a preventable cause of heart failure. Myocardial biopsy is an invasive and impractical method for patient follow-up, making it necessary to explore alternative diagnostic methods[1]. Instead, if possible, CMR should follow the T2* value[2,3]. Studies have reported that the decrease in T2* detected in CMR is associated with LV dysfunction and an increased risk of cardiac events[1-3]. T2* measurements in CMR are obtained from a single section passing through the midventricular level using a multi-echo T2* sequence on short-axis images[3,4]. The measurement is based on the change in signal intensity of the interventricular septum according to the echo time[3]. Research has shown a correlation between myocardial T2* values and myocardial iron concentration. Loading begins when this value falls below 20 ms, and the risk of developing heart failure is very high when it falls below 10 ms[2-4]. Additionally, there is a good correlation between the LV GLS value obtained from strain echocardiography and T2* values determined by CMR imaging, as reported in the literature. Therefore, in centers without access to CMR imaging, echocardiography and GLS examination can be considered as alternatives to detect subclinical dysfunction caused by iron accumulation[5].

In conclusion, iron chelation therapy should be applied to patients with TM who undergo frequent recurrent blood transfusions. Investigation of LV function and iron deposition using T2* sequence, along with echocardiographic examination including strain echocardiography and/or CMR examination, can guide the clinician in the early detection of heart failure due to secondary hemachromatosis.

Ethics

Informed Consent: Consent form was filled out by all participants.

Authorship Contributions

Surgical and Medical Practices: M.E., E.Y., Concept: M.E., M.G., Design: M.E., M.G., Data Collection or Processing: M.G., F.E.U., Y.G., Analysis or Interpretation: F.E.U., Y.G., Literature Search: M.E., E.Y., M.G., Writing: M.E., M.G.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

REFERENCES

1. Aronow WS. Management of cardiac hemochromatosis. Arch Med Sci. 2018;14:560-8.
2. Rezaeian N, Mohtasham MA, Khaleel AJ, Parnianfard N, Kasani K, Golshan R. Comparison of global strain values of myocardium in beta-thalassemia major patients with iron load using specific feature tracking in cardiac magnetic resonance imaging. Int J Cardiovasc Imaging. 2020;36:1343-9.
3. Baksi AJ, Pennell DJ. T2* imaging of the heart: methods, applications, and outcomes. Top Magn Reson Imaging. 2014;23:13-20.
4. Anderson LJ, Holden S, Davis B, Prescott E, Charrier CC, Bunce NH, et al. Cardiovascular T2-star (T2*) magnetic resonance for the early diagnosis of myocardial iron overload. Eur Heart J. 2001;22:2171-9.
5. Bonios MJ, Fountas E, Delaporta P, Kyrzopoulos S, Kattamis A, Adamopoulos SN, et al. Left ventricular deformation mechanics over time in patients with thalassemia major with and without iron overload. BMC Cardiovasc Disord. 2021;21:81.