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Case Report

Cardiac Involvement in β-Thalassemia: Prevention and Management Strategies to Prevent Fatal Progression

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Abstract: β-thalassemia is a prevalent genetic disorder characterized by chronic anemia, necessitating regular blood transfusions and resulting in iron overload, notably within the heart. This iron overload can lead to a range of cardiac complications, often culminating in heart failure, which stands as the primary cause of mortality in patients with this disorder. Early detection and timely management of these cardiac complications are paramount for ensuring the best possible outcomes for affected individuals. This article presents and discusses in detail three clinical cases, underscoring the critical importance of early screening and effective management strategies to avert fatal progression in patients afflicted by β-thalassemia.

Keywords: β-thalassemia, Cardiac involvement, Heart failure, Chelation therapy, Cardiomyopathy, Magnetic resonance imaging, Echocardiography.

INTRODUCTION

β-thalassemia, a major form of thalassemia, is a prevalent genetic disorder characterized by chronic anemia necessitating regular blood transfusions. These life-saving transfusions, however, lead to iron overload, notably within the heart, which can cause diverse cardiac complications, often culminating in heart failure. Early detection of these cardiac conditions is crucial for optimal management. This article presents three clinical cases to illustrate the necessity of early screening and management measures to prevent fatal progression in β-thalassemia patients [1, 2].

OBSERVATIONS

Case 01 (Patient A.H, 28 years)

This patient presents a complex clinical picture, including a history of type 1 diabetes, hypothyroidism, splenectomy, and blood transfusions every 21 days without chelation. She was admitted for right-sided heart failure confirmed by restrictive cardiomyopathy (CMR) as revealed by cardiac magnetic resonance imaging (MRI). Echocardiographic examination (ECHO) showed an ejection fraction (EF) of 45%, elevated ventricular filling pressures (PRVG), bi-atrial dilation, and the presence of an apical thrombus in the right ventricle (Fig 1). Unfortunately, despite medical treatment, the patient passed away a month after hospital discharge due to refractory heart failure.

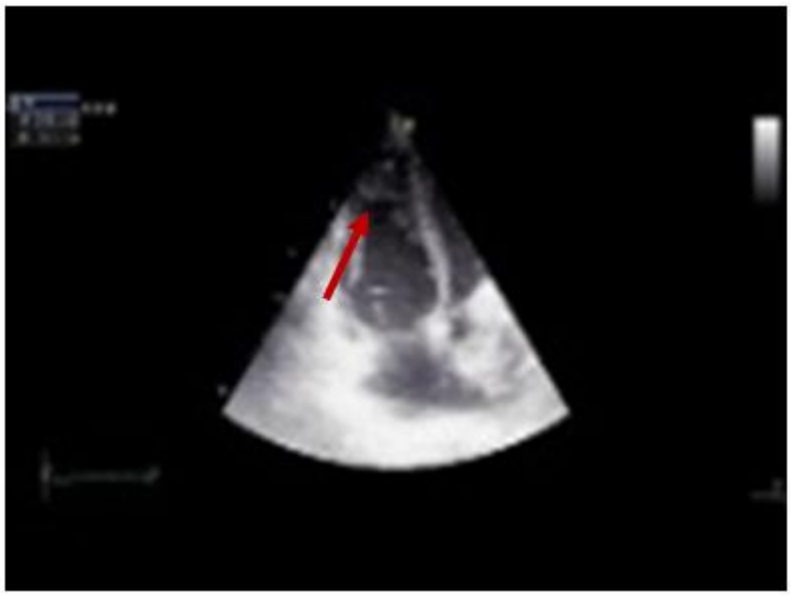

Fig. 1: Image of an echocardiography (ETT) showing a right ventricular thrombus (the arrow)

Case 02 (Patient M.M, 24 years)

This patient underwent splenectomy and has hepatic and cardiac hemochromatosis. He receives blood transfusions every 21 days and was hospitalized for overall right-sided heart failure, with dilated cardiomyopathy (DCM) complicated by atrial fibrillation (AF). The electrocardiogram (ECG) showed AF with a heart rate of 75 bpm and diffuse negative T waves. ECHO revealed an EF of 35%, altered left ventricular strain (GLS -10.5%) (Fig 2), restrictive mitral flow, biventricular dilation, left atrial dilation, and a pulmonary artery systolic pressure (PAPS) of 55 mmHg. Fortunately, with appropriate medical treatment, including bisoprolol, ramipril, spironolactone, and dapagliflozin, the patient remained stable and is being followed in cardiology consultation.

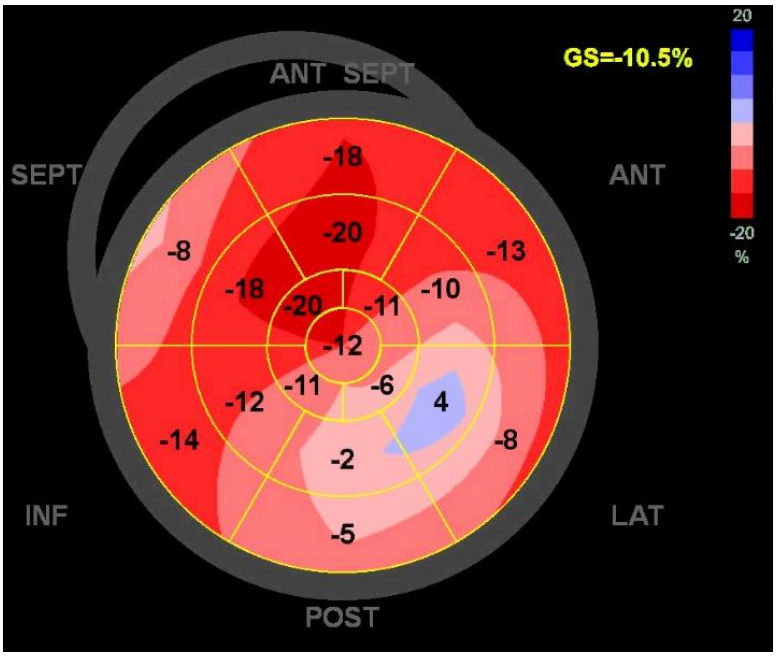

Fig. 2: Echocardiography (ETT) image showing altered left ventricular strain

Case 03 (Patient B.N, 22 years old)

Patient B.N, a 22-year-old individual, underwent splenectomy and has a medical history of complications, including an ischemic stroke, hypogonadism, an anaphylactic reaction post-transfusion, cardiac iron overload, and necessitates blood transfusions every 21 days. Hospitalization was required due to right-sided heart failure associated with confirmed restrictive cardiomyopathy (RCM) through cardiac magnetic resonance imaging (MRI). The electrocardiogram (ECG) displayed a regular sinus rhythm with multiple polymorphic ventricular premature contractions (PVCs).

Echocardiography (ECHO) revealed global hypokinesis, an ejection fraction (EF) of 38%, bi-atrial dilation, significant tricuspid regurgitation (TR), and a pulmonary artery systolic pressure (PAPS) of 40 mmHg. The patient remained stable under medical treatment, including bisoprolol, spironolactone, and furosemide, and is being regularly monitored in cardiology consultations.

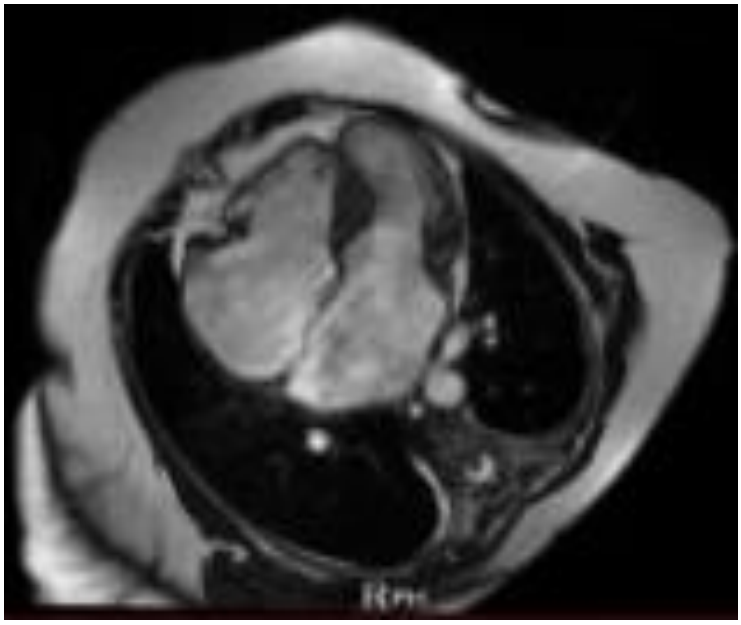

Fig. 3: Cardiac MRI appearance of restrictive cardiomyopathy (RCM)

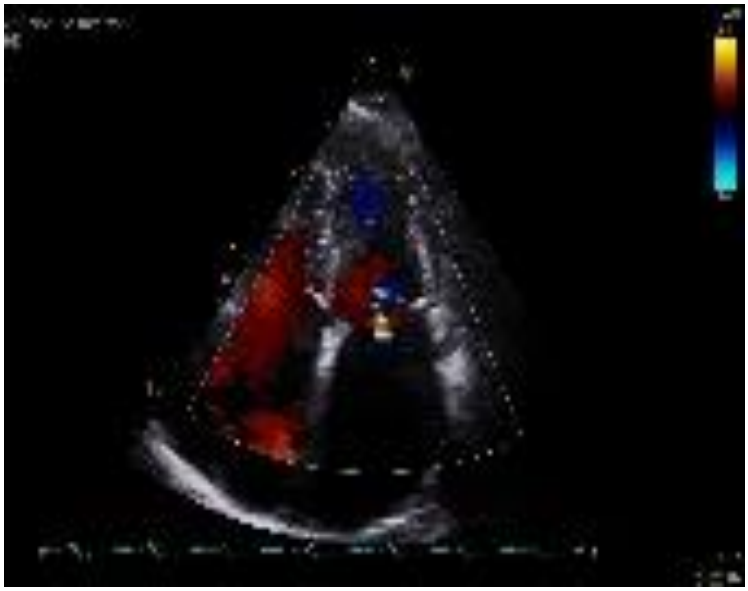

Fig. 4: Echocardiography (ETT) image depicting a typical appearance of a restrictive cardiomyopathy (RCM)

DISCUSSION

β-thalassemia is associated with a high risk of cardiac complications, particularly right-sided heart failure often accompanied by atrial fibrillation [1]. Early detection of cardiac involvement is crucial and should start at the age of 8, based on several elements:

Thorough clinical examination: Regular clinical assessments every 6 months help monitor signs of heart failure and early detection of cardiac symptoms.

- Annual ECG: ECG is a valuable tool to detect cardiac rhythm abnormalities, conduction disorders, and ventricular hypertrophy.
- Annual ECHO: ECHO is essential to detect systolic and/or diastolic dysfunction of the left ventricle or restrictive involvement, pulmonary hypertension, as well as cardiac chamber dilation and the possible presence of thrombi [2] (As in case 1).

- Cardiac MRI (T2) every 24 months: Cardiac MRI evaluates myocardial iron load, ventricular volumes and functions, and myocardial fibrosis, providing crucial information for patient management [3].

Ideally, these evaluations should be performed by a cardiologist with expertise in iron overload cardiac diseases, working closely with a hematologist in a specialized follow-up center.

If a cardiac abnormality is detected, treatment for heart failure is initiated, with medications such as bisoprolol, ramipril, and spironolactone, and close monitoring is conducted in cardiology consultations to track the disease's progression and adjust treatment accordingly. In this context, several studies have demonstrated the effectiveness of iron chelators such as deferasirox in reducing cardiac iron overload and its beneficial effects on the survival and quality of life of β-thalassemia patients [4].

This response provides an in-depth analysis of the observation from case 1, highlighting the implications of iron overload on the cardiovascular system and underscoring the importance of adequate chelation. The added references support these arguments based on research and clinical studies.

CONCLUSION

Cardiac involvement in β-thalassemia is a major complication requiring early detection and careful monitoring to prevent decompensation. Chelation therapy and standard heart failure treatment play vital roles in preventing the fatal progression of this disease. Coordinated care between specialized cardiologists and hematologists is paramount for optimal management.

Conflicts of Interest: No

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