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Acute myocarditis or apical balloon cardiomyopathy in a patient with systemic lupus erythematosus: The priority of common sense

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Abstract

Myocarditis can mimic takotsubo cardiomyopathy and via versa. We present a 70-year-old woman with known systemic lupus erythematous who presented to the emergency department twice during the last 2 years with chest pain, dyspnea and transient LV dysfunction. We also discuss the difficulties in differential diagnosis between lupus myocarditis and takotsubo cardiomyopathy.

Keywords: Systemic lupus erythematosus; SLE cardiomyopathy; Lupus myocarditis; Cardiovascular disease; Antiphospholipid syndrome.

Introduction

Systemic Lupus Erythematosus (SLE) is a chronic multi-system autoimmune connective tissue disease [1]. Cardiac involvement in SLE may concern any part of the heart: endocardium, myocardium, epicardium, pericardium, valves and coronary arteries [2-4].

This case report describes a patient with SLE who presented with cardiogenic shock, while 2 years ago she had been diagnosed as takotsubo cardiomyopathy after an episode of retrosternal chest pain and transient LV dysfunction.

Case description

A 72 year-old woman admitted to the emergency department with chest pain, dyspnea and cardiogenic shock. She had a history of SLE (diagnosed at the age of 51), antiphospholipid syndrome, pulmonary embolism, permanent atrial fibrillation, permanent VVI pacemaker and hypothyroidism. She had been treated with oral methylprednisolone (4 mg/day) and acenocumarol (target international normalised ratio: 2.5-3.0). Two years ago she had been hospitalized with chest pain after emotional stress, moderate elevated troponin and akinesia of the apical segments of left ventricle (LV). Echocardiogram showed nearly fully recovery (LVEF: 50%), and was discharged with a diagnosis of takotsubo cardiomyopathy.

On physical examination the patient was short of breath with orthopnea. She was hemodynamically unstable. Blood pressure was 75/50 mmHg and heart rate 140 beats per min (irregular rhythm). Bilateral basal crackles were heard. Electrocardiogram showed atrial fibrillation with rapid ventricular response and negative T waves in leads I, AVL, V3-V6. Chest X-ray revealed enlarged cardiac silhouette and upper lobe pulmonary venous diversion. Echocardiogram demonstrated severe LV systolic dysfunction (ejection fraction: 15%), normal LV diameters, global

hypokinesia of LV walls and high filling pressure of LV. High sensitivity cardiac troponin I was 192 pg/ml and C-reactive protein 81 mg/L. An emergent transradial coronary angiography was performed which showed coronary arteries without significant stenosis. Acute viral infection was not detected by serology tests. Complement proteins were low: C3:45.7 mg/dl, C4:6.49 mg/dl.

She received intravenous vasoconstrictors, inotropes, furosemide and high dose corticosteroids [initial dose 1 g intravenous methylprednisolone for 3 consecutive days followed by oral methylprednisolone (1 mg/kg/day)]. Further immunosuppressive medications were not started due to the presence of fever and the fear of an ongoing infection. During the next 10 days the patient showed gradual clinical and echocardiographic improvement. Serial echocardiograms showed an improvement of LV function (LV ejection fractio: 35%, end-diastolic diameter of LV 4.53 cm, normal filling pressure of the LV, estimated pulmonary artery systolic pressure 45 mmHg, normal function of the right ventricle, no pericardial fluid) she was discharged with instructions for close follow-up in the heart failure clinic and evaluation by the rheumatology department. In a follow-up visit after three months, she remained clinically stable, NYHA Class II-III and LV function in echocardiogram had no significant change. She was treated by the rheumatologists with 4 mg of methylprednisolone and azathioprine which was added one week after she was discharged from the hospital.

Discussion

Heart failure in SLE patients is usually caused by an inflammatory process within the myocardium [5]. Acute, chronic, or recurrent myocarditis is found up to 80% of autopsies [5,6]. Our patient was hospitalized twice the last two years with impaired LV function, mildly to moderately elevated troponin, low C3-C4 and no significant coronary artery disease which was excluded both times by coronary angiography. The history of SLE and antiphospholipid syndrome, the absence of physical or emotional stress, low complement proteins, the negative coronary angiography, the partially recovered LV function and the response to high dose of corticosteroids strongly supported lupus myocarditis as the most probable diagnosis [7]. The diagnosis of takotsubo cardiomyopathy in the previous hospitalization has to be challenged a posteriori. Although an emotional trigger preceded the event and nearly complete improvement of LV dysfunction suggested takotsubo cardiomyopathy [8], lupus myocarditis could not be excluded. Both syndromes have a low recurrence rate although with different outcomes [6]. LV function recovery among survivors is also frequent in lupus myocarditis [6]. The disease is often misdiagnosed while prompt diagnosis is essential since high dose corticosteroids and other immunosuppressive agents may alter prognosis and recurrence. Finally it is always important to bare in mind that takotsubo cardiomyopathy might be a manifestation of rheumatological diseases [9]. Cardiac Magnetic Resonance Imaging (MRI) is recommended to distinguish between acute myocarditis and takotsubo cardiomyopathy [8]. Considerable difference is the pattern of edema and the extent of late gadolinium enhancement. Unfortunately our patient had a non MRI compatible pacemaker. Endomyocardial biopsy is considered the gold standard for diagnosis of lupus myocarditis. Due to the fact that she was in cardiogenic shock and was under anticoagulation therapy, an emergent endomyocardial biopsy was not performed. Several case reports with left ventricular apical ballooning appeared to be caused by biopsy-proven myocarditis [10-12]. However, the method has low diagnostic sensitivity for SLE myocarditis and it is rarely performed in clinical practice.

Conclusion

In conclusion, SLE patients with acute cardiac failure not attributed in coronary artery or cardiac valve disease should be considered as lupus myocarditis until the contrary is proved. In addition patients with rheumatic conditions presented with takotsubo syndrome must always be further investigated as it might be a sign of disease activity. As a result an early intensive anti-inflammatory treatment is essential for the treatment and prognosis of the disease as wells as prevention of future events. Finally, only an MRI compatible permanent pacemaker should be implanted in SLE patients, since the use of CMI is important for evaluation suspected myocarditis, a potential life-threatening condition, in these patients.

Declarations

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