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Postpartum flare up of systemic lupus erythematosus presenting with acute decompensated heart failure mimicking postpartum cardiomyopathy

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Abstract

SLE is an autoimmune disease which often affects women during childbearing years with flare up. Endogenous estrogens are known to be the risk factor for the SLE development.This patient had the primary manifestation and diagnosis of SLE with cardiomyopathy and proteinuria as aggravating factor such as pregnancy.In a patient presenting congestive heart failure in the postpartum period,various causes of heart failure with reduced ejection fraction were always considered.

Background

Systemic lupus erythematosus (SLE) is an autoimmune and connective tissue disease characterized by the production of autoantibodies, can affect all organ systems.The heart is one of the most frequently affected organs in SLE[1, 2].

Cardiac involvement presents in up to 50% of SLE patients and pericarditis is the most frequent manifestation of SLE-related cardiac disease[3]. However, all other cardiac components may be involved: endocardium, myocardium, conduction tissue, and coronary arteries[2]. Myocardium involvement of SLE is a rare but potentially fatal complication, affecting up to 10% of SLE patients[3, 4]. It may present as an acute illness or have a chronic course with the development of cardiomyopathy.

If the underlying disease of patient was not confirmed, the first suspicion about left ventricular dysfunction after delivery could be postpartum cardiomyopathy. In a patient presenting left ventricular systolic dysfunction with heart failure in the postpartum period, we would like to discuss that this condition would have various etiologies and might be difficult to diagnose.

Case report

A 33-year-old pregnant woman underwent a caesarean section at 38 weeks. After delivery about 4-5 days, this patient complaint of orthopnea, serious pitting edema, and overt weight gain. This patient was transferred to the emergency room of our hospital. Pulmonary edema and cardiomegaly were confirmed on chest radiography (figure 1), and left ventricular mild dysfunction (ejection fraction about 43%) and global hypokinesia was detected on initial echocardiography (video 1). The clinician suspected that this patient's condition was diagnosed of postpartum cardiomyopathy and attempted to control the congestion using diuretics. Initial vital sign revealed slight hypertensive blood pressure (169/100mmHg – 90bpm), and then after using diuretics, slightly controlled blood pressure was detected (134/80mmHg – 83bpm).

However, the symptom as dyspnea and orthopnea was worsened, and fever (38.3C`) was occurred. It has been about 10 days since using diuretics, however, there was no improvement in congestion and the O2 demand was rather increased (figure 2), and ejection fraction deteriorated (about 30%) in followed up echocardiography (video 2). As considering that there may be other causes, the patient's past history was reviewed.

We confirmed that proteinuria 3+, neutropenia and thrombocytopenia occurred on 3 months before delivery. Proteinuria (protein/creatinine ratio 3133.2mg/g) continued while using diuretics after delivery, which additionally we suspected another cause as glomerulonephritis. The lowering complements level (C3 40mg/dL, C4 3mg/dL) was confirmed. Finally, ANA (1:320 Speckled pattern) and Anti-dsDNA Ab (168IU/mL) were additionally confirmed as positive, and SLE was diagnosed. After SLE was diagnosed, congestion was slightlycontrolled by using steroids.

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Discussion

SLE cardiomyopathy is a severe cardiac manifestation of SLE, treated with high-dose corticosteroids, with or without other immunosuppressive therapy, in addition to standard cardiac management[5, 6]. Because SLE-related cardiomyopathy is rare, there are few prospective studies, and management is based on isolated cases or small-series reports.Up to 9% of SLE patients are reported to have cardiac manifestations including myocarditis, cardiomyopathy, and/or heart failure.

Myocardial inflammation may not be the pathophysiological basis for the acute myocardial decompensated seen in SLE patients, and that several other acute and active factors such as: fever, infection, anemia, uremia, hypertension, and accelerated atherosclerosis, may play a role in the development of SLE cardiomyopathy though various physiologic mechanisms causing impaired cardiovascular function and resulting in a process such as stress-related cardiomyopathy.

The diagnosis of SLE cardiomyopathyremains challenging in clinical practice. Cardiac magnetic resonance (CMR) imaging is a sensitive noninvasive technique to investigate myocarditis. However, CMR alone is insufficient to clarify the cause of cardiomyopathy. Although endomyocardial biopsy is considered to be the gold standard for the diagnosis of myocarditis, it is not performed routinely because endocardial biopsy is invasive and has a risk of possible sampling error. Currently, the most reasonable strategy in clinical practice is considering the medical history combined with a positive laboratory test when SLE is suspected[7].

Estrogen possesses both immune-stimulating and immunosuppressive properties[1, 4, 8].SLE is an autoimmune disease which often affects women during childbearing years with flare up. In some situations, exogenous estrogen predisposes to development of new SLE, flares of preexisting SLE, and thromboses in susceptible individuals.

This patient had the primary manifestation and diagnosis of SLE with cardiomyopathy and proteinuria as aggravating factor such as pregnancy. Because we focused on the postpartum condition, various causes of heart failure with reduced ejection fraction were not considered from the beginning.

We always consider the most probable cases for searching the cause of heart failure. If there is no adequate response to general and usual treatment, various other causes must be considered. This patient also mimicking postpartum cardiomyopathy as SLE presenting with disease flare up and heart failure.

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