La myocardite lupique : A propos de deux cas

Myocarditis complicating systemic lupus erythematosus: two case reports

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Résumé

Introduction : La myocardite est une manifestation rare et grave du lupus érythémateux systémique (LED). La présentation clinique et le pronostic sont variables.
Cas cliniques : Nous rapportons les cas de deux patients suivis pour LED qui ont présenté un tableau clinique d'insuffisance cardiaque. Le diagnostic de myocardite lupique a été retenu en se basant
Conclusion : Bien que la myocardite soit une manifestation rare du LES, elle peut être une maladie potentiellement mortelle.

Mots-clés
Lupus érythémateux systémique, myocardite, insuffisance cardiaque

Summary

Background: Moycarditis is a rare and serious manifestation of systemic lupus erythematosus (SLE), with numerous clinical presentations and variable outcome.
Case reports: We report the cases of two patients with SLE who presented with clinical signs of heart failure, the diagnosis of lupic myocarditis was retained. The clinical features and management will be discussed.
Conclusion: Although myocarditis is a rare manifestation of SLE, it may be a life-threatening condition.

Keywords
Systemic lupus erythematosus, myocarditis, heart failure

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INTRODUCTION

Systemic lupus erythematosus is a chronic, complex autoimmune connective-tissue disorder of unknown etiology. It can affect all organ systems with significant clinical and immunological diversity. Lupus myocarditis is an uncommon and life-threatening complication of SLE [1].

Here, we report the cases of two patients presenting with lupicmyocarditis.

CASE REPORTS

Patient 1

A 32-years old woman with known SLE, presented acutely with 24 hours history of dyspnea, central chest pain and palpitation.

The diagnosis of SLE was retained three years ago based on ACR (American college of Rheumatology) criteria: arthralgia,malar rash, photo sensibility and positive antinuclear antibodies. She was put on hydrochloroquine and low dose steroids.

Physical examination showed: fever of 38° C, tachypnea with a respiratory rate of 28 breaths per minute, tachycardia with a heart rate of 100 beats per minute, blood pressure at 120/70 mmHg, and she had crepitantrales at bases of both her lungs. The electrocardiogram (ECG) showed sinus rhythm with ventricular extra systoles. Chest X -rays demonstrated bilateral interstitial and alveolar edema.

Laboratory findings revealed a CRP: 40 mg/dl, normochromic anemia with hemoglobin at 9.5 g/dl, elevated troponin level, the immunological workup showed increased anti-DNA antibodies, decreased levels of complement C3/C4 and positive anti-Sm antibodies.

Two-dimensional transthoracic echocardiography (TTE) revealed left ventricular systolic function impairment with global hypokinesis and left ventricular ejection fraction (LVEF) of 33%, along with small pericardial effusion. A CT scan was performed and showed no significant coronary artery stenosis.

Cardiac MRI showed delayed gadolinium enhancement (DGE) in a non-vascular pattern, and confirmed LV dysfunction (Figure 1).

The diagnosis of acute lupic myocarditis was retained.

The patient was treated with intravenous corticosteroids at a dose of (1 mg/kg) for three days and azathioprine 150 mg/day, in association with the usual treatment of heart failure.

After one week, the patient had clinical improvement. Control echocardiography performed one month later, showed improvement of the LVEF to 51%.

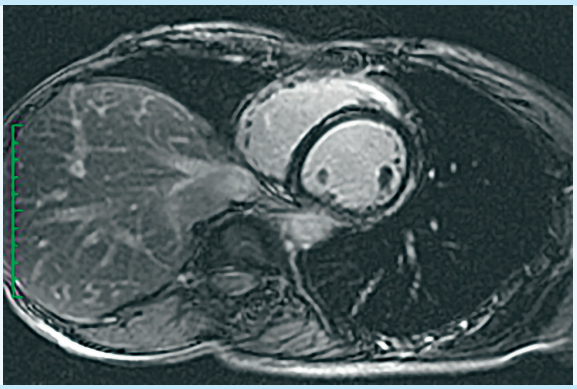

Figure 1: Cardiac MRI showing delayed gadolinium enhancement (DGE) in a non-vascular pattern.

Patient 2

A 21-year-old woman diagnosed with SLE, presented after two years of follow-up, with 72-hour history of dyspnea, pedal edema and orthopnea.

The physical examination revealed fever with a temperature of 38,4 °C, the respiratory rate was 20 breaths per minute, and the cardiac rate was110 beats per minute. The blood pressure was at 110/70 mmHg.

Cardiac examination revealed normal heart sounds and no murmur, pulmonary examination revealed bilateral basal crackles. Lower limb edema was also noted.

The ECG showed a sinus tachycardia with negative T waves in the anterior leads.

A chest radiograph demonstrated cardiomegaly and interstitial edema suggestive of acute pulmonary edema.

Biology revealed elevated troponin and pro BNP level. Complete blood count, Renal and liver function test results were normal, CRP was at 90 mg/l.

The immunological workup showed a high positive ANA titer, positive anti-double stranded DNA antibodies, decreased complements level, and positive anti-Ro/La antibodies.

The Echocardiography showed biventricular dilatation with left ventricular systolic function impairment with global hypokinesis, LVEF was 25%; pulmonary artery systolic pressure (PASP) was 40mmHg.

CT scan demonstrated normal coronary arteries.

Cardiac MRI revealed radiological patterns of myocarditis and confirmed the dysfunction of the left ventricle, the LVEF was 20%.

The diagnosis of acute myocarditis secondary to SLE was made, based on the clinical findings and investigations.

Remission of the cardiac signs and symptoms was obtained under corticosteroids (1mg/kg), immunosuppressive treatment (azathioprine) and usual heart failure treatment.

LVEF was controlled three months later and showed improvement of left ventricular systolic function the LVEF was 48%.

DISCUSSION

Heart involvement is observed in more than 50% of patients with SLE [2]. All the cardiac structures may be involved.

Though clinical myocardial involvement is infrequent in SLE, it is noticed in up to 50-80% of cases on necropsy studies [3]. These findings suggest that the majority of cases are subclinical [3].

Indeed, in a prospective study of 70 patients, echocardiography revealed myocardial abnormalities in 14 cases (20%) yet solely one patient was symptomatic [4].

The prevalence of symptomatic LM has been assessed up to 9% in other series. It can occur any time during SLE evolution [5].

Lupic myocarditis is a life-threatening condition that may have heterogenous clinical features such as dyspnea, fever, chest pain, conduction disturbances, arrythmias and heart failure [5]. It can rarely be the revealing presentation of SLE [6].

Laboratory findings are nonspecific and may include elevation of troponin as presented by our patients [5].

Endomyocardial biopsy remains the gold standard for diagnosing lupus myocarditis. Yet, it has a poor sensitivity and specificity because of the patchy myocardial involvement. In addition, it has a high risk of complications [7].

Hence, the diagnosis confirmation is based on clinical findings, biological markers, and echocardiography [5].

Cardiac MRI is a safe procedure that can give tissue characterization, differentiate between inflammatory and ischemic lesions. Thus, MRI can detect LM with a high degree of diagnostic precision [8].

Myocarditis is a rare and serious complication of SLE. Subsequently, there are no randomized trials on treatment strategy.

Limited evidence exists regarding optimal treatment and prognosis; thus, management is based on small series or isolated case reports.

High dose corticosteroid is the most common prescribed treatment [9], but the duration, doses, pulses and rhythm of tapering are not well determined [8].

Immunosuppressive therapy has also been used in LM, with significant improvement of the systolic function especially in patients with more severe and lower EF [10].

Other reports suggest that the biological agents may have encouraging results and can be useful for the management of LM [9].

Rituximab has also been successfully used in a pediatric case[10].

The long-term outcome is rarely reported in the literature.

Accordingly, Thomas and al. showed in a multicentric retrospective study including 29 patients that the long-term prognosis was favorable with a mortality rate up to 7% [10]. The median LVEF was 37% at baseline, all the patients had an LVEF over than 50% at the end of follow up which suggest that LM is severe at the beginning.

Both our two patients had an improvement of the left ventricular systolic function.

CONCLUSION

Lupus myocarditis is a rare, but potentially fatal complication of systemic lupus erythematosus (SLE). Here, we report the initial course and outcome in two patients with SLE who developed left ventricular failure secondary to lupicmyocarditis.

Larger series are needed to further describe the demographic, clinical, immunological features of LM and to guide therapeutic management.

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