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Short Article

Long-term successfull treatment of patient with history of Systemic Lupus Erythematosus complicated with Acute Myocarditis and Dilated Cardiomyopathy – case report

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ABSTRACT

Introduction and aim. Systemic lupus erythematosus (SLE) is a chronic, autoimmune disorder that may affect every organ. The use of immunosuppression has changed a natural course of the disease. Thereupon mortality in SLE is not only associated with implications of disease activity and its impact on cardiovascular system but also with treatment-related complications like cytopenia or increased risk of infection, which can lead to a life-threatening sepsis.

Description of the case. We present a case of a 60-year-old Caucasian female patient with a long-time history of active SLE, who has first developed nephrotic syndrome and then acute lupus myocarditis (LM) complicated by dilated cardiomyopathy.

Conclusion: The appearance of LM and its complications seem to be a landmark event in the further course of disease.

Key words. systemic lupus erythematosus, lupus myocarditis, lupus nephritis, dilated cardiomyopathy, cardiorenal syndrome

Introduction

Dilated cardiomyopathy is an uncommon complication of LM which can be secondary to SLE. Symptomatic left ventricle dysfunction is a life-threatening condition and it is the most frequent presentation of cardiomyopathy. Not only is heart failure a reason why acute LM require immediate medical care, but also there is a risk of development severe complications such as cardiac arrhythmias.[1] We have decided to describe this case, because mostly mortality in SLE is rarely related to myocardial involvement.[2] Besides majority of patients with SLE complications in the form of LM presents at some point apparent improvement of heart function, like our patient did, but soon she developed very uncommon complication – dilated cardiomyopathy, which has significant impact on a course of disease.

Case report

A 61-year-old woman with a twenty-one-year history of SLE presented to the emergency department of our hospital with an exacerbation of chronic heart failure. She complained of dyspnoea at rest, fatigue and increasing oedema of lower legs since 3 days. Patient's heart failure symptoms were classified as class IV according to New York Heart Association (NYHA) classification. At admission her general condition was serious, but she maintained logical verbal contact and did not report stenocardial complaints. Moreover, the patient had a history of chronic coronary syndrome, hypertension, chronic obstructive pulmonary disease, diabetes mellitus type 2,

severe chronic renal failure, thrombosis of the left popliteal vein in the past, anemia of chronic disease. In physical examination: massive oedema of legs and face, dilated jugular veins, heart rate -regular, 100 beats per minute, blood pressure - 150/100 mmHg, respiratory rate - 36 breaths per minte, oxygen saturation - 90% on room air, dull percussion note over the right lung, redness and pain of left breast. In laboratory findings: NT-proBNP - 324 060 pq/ml (N<226 pg/ml), troponin T - 0,686 ng/ml (N <0,014 ng/ml), creatinine - 4,92 mg/dl (N <0,90 mg/dl), C-reactive protein - 267 mg/l (N<5 mg/l), hypoalbuminemia - 18g/l (N:35-50 g/l). Chest radiography revealed fluid in right pleural cavity up to a level of fourth rib. Echocardiography disclosed global left and right ventricular hypokinesia with reduced ejection fraction of left ventricule (LVEF = 30 %), decreased right ventricular global systolic function – tricuspid annular plane systoli excursion (TAPSE = 15 mm) and left ventricular enlargement. Echocardiography showed left bundle branch block (LBBB). Performed breast ultrasound revealed inflammatory changes without enlarged lymph nodes. Patient chronically received cyclosporine and methylprednisolone. (Fig. 1) During hospitalization, despite intensive pharmacological cardiovascular treatment and renal replacement therapy, features of heart failure were strengthened by sepsis and after all the patient died.

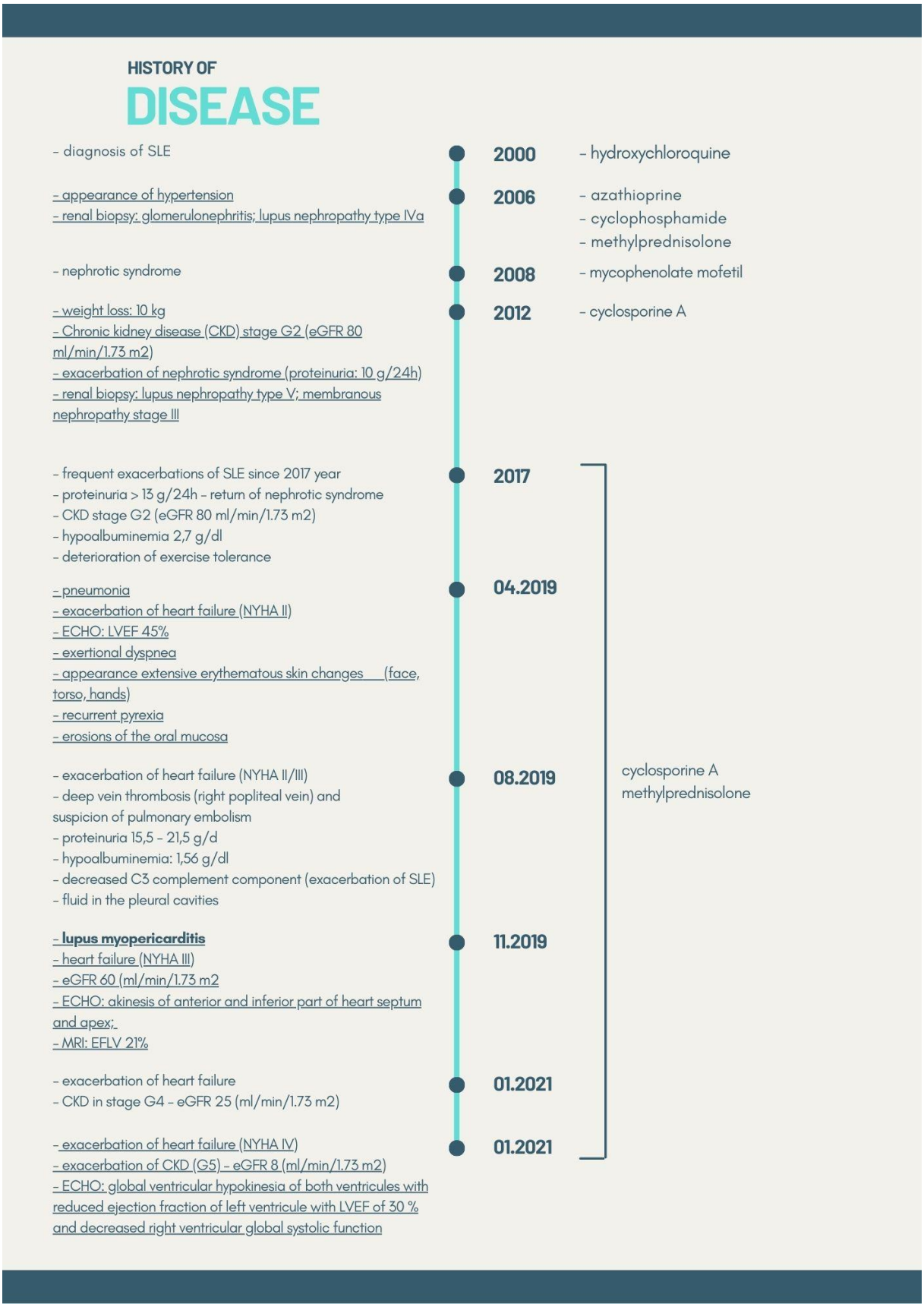

Figure 1 - SLE course and immunosuppressive treatment in our patient over the years

Discussion

We describe a case of the patient with over twenty-year-history of SLE, who presented with symptoms of post-inflammatory dilated cardiomyopathy 15 months after diagnosis of LM. The diagnosis of LM in our patient is based on clinical manifestations of left ventricle insufficiency, confirmed echocardiographically (significantly decreased LVEF and global hypokinesia of myocardium). The long-term outcomes of LM are variable and based mostly on individual case reports, additionally in majority at some point LVEF improves and the recurrences are rare. Sometimes, LM may lead to development of dangerous complications such as dilated cardiomyopathy, heart failure or cardiac arrhytmias.[2] SLE is primarily diagnosed in young age, with the peak incidence falling on the 29th year of life. Our patient was diagnosed with the disease at the age of 40. Population-based cohort studies have shown that the ten-year survival rate in incident cohorts span from 90% in Europe, to 93% in recent study from Norway.[3]

Mortality in SLE is almost 2.6-fold greater when compared to age- and sex-matched comparisons.[4] Reasons of death in SLE patients are usually grouped according to clinical manifestations in three most common situations: underlying disease activity, cardiovascular complications and infections. Cardiovascular disease was the most frequent cause of death in studies with follow-up times over 15 years.[5] When lupus nephritis develops, the risk of mortality increases 6-fold in comparison with the general population. In case of a complication in the form of SLE-related end-stage renal disease, it comes to 26-fold excess in a risk of death.[6] This complication occurs in 5–10% of patients with SLE.[5,7] There are many other factors associated with an increased risk of mortality in SLE patients, including an early onset of SLE, degree of organs damage, comorbidities: infections, hypertension, dyslipidemia, arteriosclerosis, coronary arterial disease, diabetes mellitus, osteoporosis and cancers (according to European Alliance of Associations for Rheumatology – EULAR)[8,9] and applying high-dose glycocorticosteroids.[3] Patients who survived to later stages of SLE can develop serious complications, not directly related to underlying disease, but associated with applied treatment. Nonselective immunosuppressive therapy and severe infections are the leading causes of mortality in SLE patients.[6] In our case, mentioned above risk factors overlapped, which worsened the course of disease. Correct maintenance therapy of major organ involvement in SLE is similar, and consists of systemic glycocorticosteroids, most along with immunosuppressive agents and also supportive cardiac therapy in case of heart failure (angiotensin-converting enzyme inhibitors, beta-blockers and diuretics), and such treatment was applied in the case of our patient.[2] (Fig. 1)

According to Zhang et all 80% of patients with LM, after follow-up (mean duration -15 months), improved LVEF or showed complete recovery in ventricular dilation. Only 20% of patients in this study suffered from deterioration in wall motion abnormalities and symptoms of heart failure[1]. 15 months after onset of LM, our patient had presented ventricular hypokinesia in echocardiography and LVEF = 30%. It is impossible not to mention about the impact of increasing renal failure on development of cardiomyopathy called cardiorenal syndrome. Progressive renal failure is considered to be an independent risk factor of cardiovascular diseases. In patients with SLE underlying pathology, as well as used treatment, has influence on development of renal failure. It should be noted, that our patient suffered from type 2 diabetes and hypertension, which could impair kidney function. In our case, we could observe renal failure up to its end stage, which is likely to had impact on cardiomyopathy evolution. At the end our patient developed sepsis, which exacerbated multi-organ failure, including heart and kidneys, and finally led to death. According to Appenzeller et all death in SLE was mainly not related to myocardial involvement.[2]

Conclusion

In conclusion, LM and its complication in the form of dilated cardiomyopathy is relatively rare, but potentially fatal manifestation of disease. Comorbidities, such as diabetes mellitus, hypertension and renal insufficiency may have adverse impact on prognosis. However, quick recognition and appropriate using of supportive and immunosuppression treatment usually result in a favorable course of disease and enable to increase survival time, even in the case of severe complications of SLE.

References

1. Zhang L, Zhu YL, Li MT, et al. Lupus Myocarditis: A Case-Control Study from China. *Chin Med J (Engl)*. 2015;128(19):2588-2594.

2. Appenzeller S, Pineau CA, Clarke AE. Acute lupus myocarditis: Clinical features and outcome. *Lupus*. 2011;20(9):981-988. doi:10.1177/0961203310395800

3. Bultink IEM, de Vries F, van Vollenhoven RF, Lalmohamed A. Mortality, causes of death and influence of medication use in patients with systemic lupus erythematosus vs matched controls. *Rheumatology (Oxford)*. 2021;60(1):207-216.

4. Ocampo-Piraquive V, Nieto-Aristizábal I, Cañas CA, Tobón GJ. Mortality in systemic lupus erythematosus: causes, predictors and interventions. *Expert Rev Clin Immunol*. 2018;14(12):1043-1053.

5. Reppe Moe S, Haukeland H, Molberg Ø, Lerang K. Long-Term Outcome in Systemic Lupus Erythematosus; Knowledge from Population-Based Cohorts. *J Clin Med*. 2021;10(19):4306. Published 2021 Sep 22.

6. Trager J, Ward MM. Mortality and causes of death in systemic lupus erythematosus. *Curr Opin Rheumatol*. 2001;13(5):345-351.

7. Yap DY, Tang CS, Ma MK, Lam MF, Chan TM. Survival analysis and causes of mortality in patients with lupus nephritis. *Nephrol Dial Transplant*. 2012;27(8):3248-3254.

8. Smolen JS, Landewé RBM, Bergstra SA, et al. EULAR recommendations for the management of rheumatoid arthritis with synthetic and biological disease-modifying antirheumatic drugs: 2022 update [published correction appears in Ann Rheum Dis. 2023 Mar;82(3):e76. doi: 10.1136/ard-2022-223356corr1]. *Ann Rheum Dis*. 2023;82(1):3-18. doi:10.1136/ard-2022-223356 ‘

9. Fanouriakis A, Kostopoulou M, Andersen J, et al. EULAR recommendations for the management of systemic lupus erythematosus: 2023 update. *Ann Rheum Dis*. 2024;83(1):15-29. Published 2024 Jan 2. doi:10.1136/ard-2023-224762