Massive pericardial effusion – An uncommon initial presentation of systemic lupus erythematosus (SLE)

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SUMMARY

Systemic lupus erythematosus (SLE) is a common autoimmune disease that we see in our daily clinical practice. It can involve almost every organs in the body. Cardiac manifestations of SLE include pericarditis, myocarditis, heart block, coronary artery disease and others. Here, we report a case of SLE with an uncommon presentation of massive pericardial effusion as initial presentation. Here we also highlight that massive pericardial effusion can also be associated with other complications of SLE such as heart failure and lupus nephritis.

INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune disorder that can affect virtually every organ in the body. Its prevalence is estimated to be around 15-50 per 100,000 individuals with female predilection. Pericarditis is a common complication of the disease but it is generally uncommon to have large volume effusion as initial presentation. When it does occur, SLE is usually associated with other complications such as nephrotic syndrome, heart failure or constrictive pericarditis. Here, we reported a case of SLE with massive pericardial effusion as initial presentation.

CASE REPORT

A 22-year-old woman with no past illness presented with bilateral leg swelling, fever and non-productive cough for two weeks. She also complained of abdominal distension and decreased effort tolerance for two days. Otherwise, she reported no joint pain, malar rash, photosensitivity, chest pain, or dyspnoea. Upon examination, her blood pressure was 128/94mmHg, pulse rate was 78 beats per minute, Spo2 was 100% on air. There was absence of Beck's triad or Kussmaul sign. Her lung examination was unremarkable. Her abdomen was distended with ascites. There was bipedal oedema. Her haemoglobin level was 7.2g/dl, total white blood cell was $3.7x10^9$/L, platelet was 174 $x10^9$/L. Renal profile was normal. Albumin was 22g/L. Urine examination showed proteinuria of 3+, erythrocyte of 1+. 24 hour urine protein was 2.5g/day. Electrocardiogram showed sinus tachycardia, no electrical alternans. Chest x-ray showed bilateral pleural effusion with cardiomegaly. Subsequent workup confirmed her diagnosis of SLE with positive antinuclear antibodies (ANA), 1:320, homogenous pattern and hypocomplemenemia. Echocardiogram showed that she had a massive global pericardial effusion, averaging 2-3cm, associated with reduced ejection fraction of 47%. She was treated with intravenous hydrocortisone. Repeated echocardiogram two weeks later showed similar findings. She was then given IV methylprednisolone 500mg OD for 3 days and cyclophosphamide 750mg. One month after cyclophosphamide, her pericardial effusion persisted. She then underwent pericardiocentesis. She eventually completed six cycles of cyclophosphamide and is keeping well since then. Repeated echocardiography at the end six cycles showed marked improvement with no pericardial effusion.

DISCUSSION

SLE is a systemic autoimmune disorder that can affect virtually all organs in the body. It is well known to have high mortality and morbidity without appropriate therapy. In the past, diagnosis of SLE is based on 1997 American College of Rheumatology criteria. SLICC criteria was introduced in 2012 to enable earlier diagnosis of SLE. SLICC criteria requires either meeting ≥4 of 17 criteria, including at least one clinical and one immunological criteria, or demonstrating biopsy-proven lupus nephritis with positive ANA or anti double-stranded (dsDNA.[1] Our patient fulfilled both ACR and SLICC criteria and she had positive ANA, hypocomplemenemia, serositis, lymphopenia, nephrotic syndrome and alopecia.

Cardiac diseases affect 15-50% of SLE patients. Cardiac manifestation in SLE patients includes pericariditis, myocarditis, endocarditis and conduction system abnormalities.[2] Pericarditis is the commonest of all, occurring in 75% of patients. Although serositis and pericarditis are common initial presentation of SLE, it is rather uncommon to have large volume effusion to occur as initial presentation. When it occurs, it is usually associated with more aggressive disease and other complications of SLE such as heart failure, constrictive pericarditis, nephrotic syndrome or Budd Chiari syndrome.[3] In our patient, she had massive pericardial effusion with heart failure and biopsy proven class IV lupus nephritis.

Regarding pathophysiology it is believed in SLE immune complex plays a major role. Granular deposition of immunoglobulin and C3, demonstrated by direct immunofluorescence, support the role of immune complexes in the development of pericarditis.[4] In SLE, pericardial effusion is usually diffuse, however, it can also be loculated

This article was accepted: 1 June 2019
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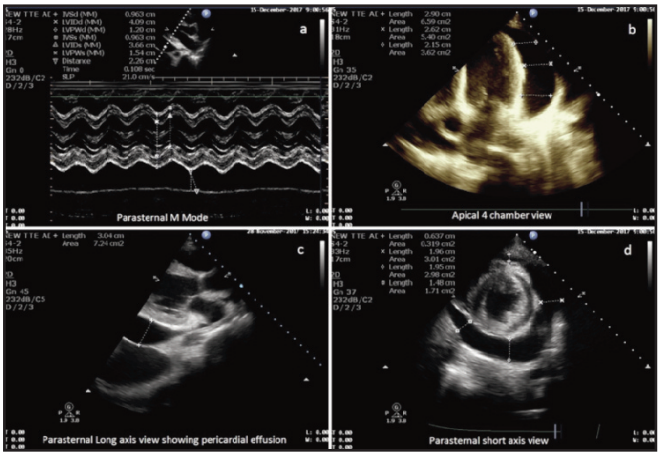

Fig. 1: Echocardiography Images of the patient.

pericardial effusion. The effusion is usually exudative in nature. In approaching a SLE patient with pericardial effusion, the other differential diagnosis to be considered include viral, bacterial, tuberculous, uremic, post myocardial infarction, idiopathic, etc. Diagnosis of pericardial effusion can be established with echocardiography.

Regarding treatment, pericarditis and pericardial effusion in SLE generally respond well to glucocorticoid and seldom require aggressive immunosuppressants such as cyclophosphamide. However, in our patient did not respond to the usual therapy of high dose steroid. Even with cyclophosphamide, her effusion did not improve and eventually needing a pericardiocentesis to relieve the effusion. It is postulated in reports that failure of therapy could be due to high levels of cytokines in pericardial fluid which impede the delivery of glucocorticoid and cyclophosphamide to the pericardial cavity.[5]

CONCLUSION

In the past, Dubois et al., reported that lupus serositis does not cause significant effusions. However, more reports have been published suggesting that massive lupus serositis does occur and can be the main initial manifestation of SLE. When it occurs, it usually signifies more active disease and is thus associated with other complications such as heart failure and nephrotic syndrome as illustrated in our case. Our case emphasizes the importance of accurate and prompt diagnosis of SLE and the early initiation of treatment resulted in the recovery of our young patient.

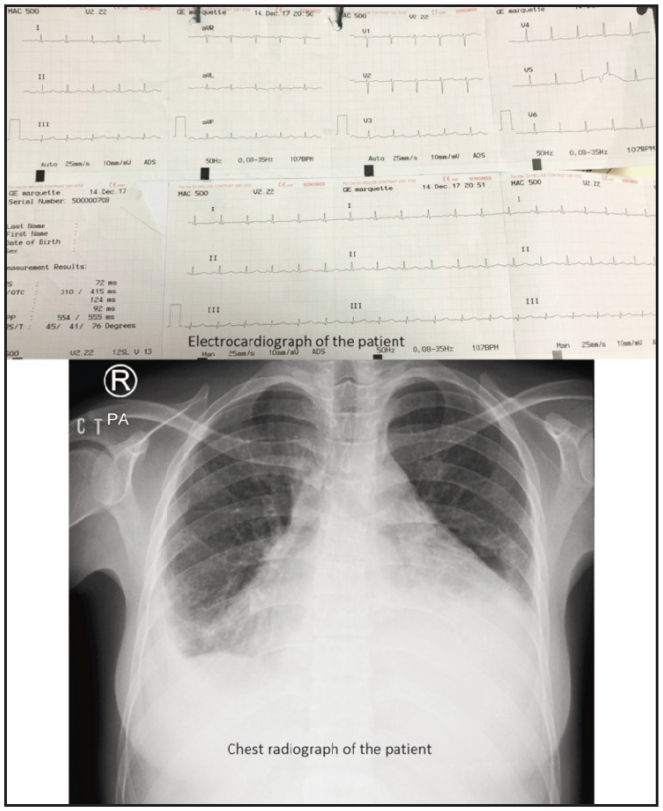

Fig. 2: A) Electrocardiograph of the patient.
B) Chest Radiograph of the patient.

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