

Systemic Lupus Erythematosus Initially Manifesting as Acute Pericarditis Complicating With Cardiac Tamponade : A Case Report

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Abstract

A 23-year-old woman was admitted with progressive shortness of breath. Echocardiography showed a large volume of pericardial effusion, which indicated cardiac tamponade. Yellowish and puriform fluid with increased white blood cell count (neutrophil dominant) was aspirated, but antibiotics were ineffective. Further examination revealed the presence of positive anti ds-DNA antibody, anti SS-A antibody and anti Sm antibody, resulting in a diagnosis of systemic lupus erythematosus. Her condition was smoothly improved by prednisolone administration. Cardiac tamponade is a rare initial manifestation of systemic lupus erythematosus.

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Key Words

- Cardiac tamponade
- Complications (systemic lupus erythematosus)
- Pericarditis (neutrophil dominant pericardial effusion, steroids reactive)

INTRODUCTION

Serositis including pericarditis is a common manifestation of systemic lupus erythematosus (SLE), and pericarditis is the most common cardiac manifestation of this condition.¹⁾ Cardiac tamponade can occur during any stage of SLE pericarditis, but rarely appears as the initial manifestation of serositis.²⁾ Moreover, cardiac tamponade is extremely rare as the initial manifestation of SLE.³⁾ This report describes a case of SLE initially manifesting as cardiac tamponade.

CASE REPORT

A 23-year-old Japanese woman with a history of hyperthyroidosis and gestosis was admitted to Jichi Medical School Hospital with a 1-month history of fever and cough. Her body temperature had increased to 38.5°C 2 days prior to admission, and was resistant to several antibiotics. Chest radiography showed purse string-like cardiac appearance, and ultrasonography revealed remarkable pericardial effusion (Figs. 1, 2).

The patient had no history of diseases associated with cardiac tamponade, such as aneurysm, myocardial infarction, amyloidosis, trauma, or

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Fig. 1 Chest radiograph showing purse string-like cardiac appearance

renal failure.⁴⁾ She also had no symptoms of SLE prior to admission, such as photosensitivity, arthritis, skin rash and neuropathy. Her height was 162 cm, weight 48 kg, blood pressure 80/60 mmHg, pulse rate 122 beats/min, body temperature 37.2°C, and her thyroid gland was not palpable. No edema was found in the arms and legs.

Electrocardiography revealed sinus tachycardia and ST elevation in all leads except for aVR. Laboratory tests upon admission showed white blood cell count 8,500/ μ l, hemoglobin 12.9 g/dl, platelet count 329,000/ μ l, serum creatinine 0.84 mg/dl, serum total protein 8.3 mg/dl, albumin 3.1 mg/dl, ALT 67 U/l, AST 114 U/l, LDH 334 U/l, CPK 49 U/l, CRP 6.22 mg/dl, Na 126 mmol/l, K

5.0 mmol/l, Cl 95 mmol/l, prothrombin time 14.7 sec, international normalized ratio 1.35, activated partial thromboplastin time 42.3 sec, C3 76 mg/dl, C4 9 mg/dl, CH50 26.9 U/ml. Echocardiography revealed a large volume of pericardial effusion, with ejection fraction of 60%, and dilatation with poor collapse of the inferior vena cava. Valvular function was normal.

On admission, she exhibited progressive shortness of breath and a markedly distended neck vein. Immediate pericardiocentesis aspirated 225 ml of yellowish and puriform fluid with opacity. The opacity solidified and precipitated at the bottom of a test tube (**Fig. 3**). Pericardial fluid analysis revealed neutrophil dominant white blood cell count 22,100/ μ l (neutrophil 81.5%, monocyte 14.0%, lymphocyte 4.5%), characteristic of a bacterial infection. Imipenem/cilastatin sodium was prescribed. On the following day, echocardiography showed almost the same volume of pericardial effusion as the previous day. No bacteria were observed in the fluid.

Three days after admission, antinuclear antibody (ANA) and rheumatoid arthritis particle agglutination test were positive, indicating the possibility of connective tissue disease, which was confirmed by the presence of anti double stranded (ds)-DNA antibody, anti SS-A antibody and anti Sm antibody. Therefore, the cause of the pericarditis was identified as SLE. Prednisolone 30 mg/day was administered and her pericarditis improved soon.

The diagnosis of SLE was established on the presence of serositis (pericarditis), leukopenia and detection of ANA, anti ds-DNA antibody, anti SS-A antibody and anti Sm antibody matching 4 of the

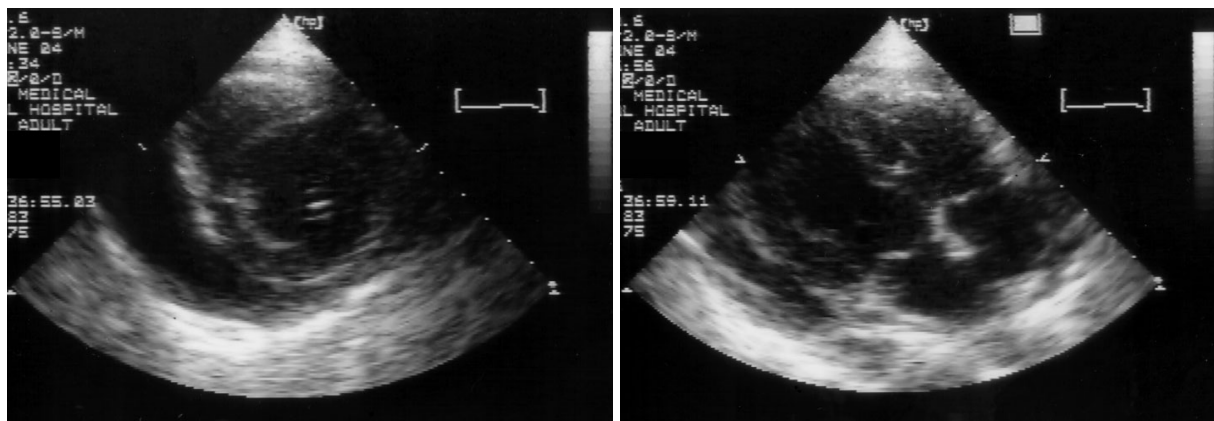


Fig. 2 Cardiac ultrasound images showing pericardial effusion
Left: Short-axis view. Right: Long-axis view.



Fig. 3 Photograph of the product of pericardiocentesis

Puriform deposit was observed at the bottom of the yellowish fluid. Neutrophil dominant white blood cell increase seemed to indicate bacterial infection.

11 SLE American Rheumatology Association (ARA) criteria for SLE. Follow-up echocardiography revealed a small amount of pericardial effusion. One month after admission, the patient was discharged.

DISCUSSION

This report describes a case of SLE which initially manifested as cardiac tamponade. The cellular content of the fluids sampled from SLE patients is usually yellowish, exudative and rarely bloody.⁵⁾ Most patients exhibit a preponderance of neutrophils with some fluids containing up to 96% neutrophils.⁶⁾ In our patient, the pericardial fluid showed a white blood cell count of 22,100/ μ l (neutrophil 81.5%, monocyte 14.0%, lymphocyte 4.5%) that is consistent with SLE. However, the fluid was puriform with opacity, suggesting bacterial infection, although the patient's history showed no events associated with bacterial pericarditis such as trauma, operation, or compromised host. Nevertheless, treatment by antibiotics was initially chosen. However, a culture of the pericardial fluid revealed no bacterial infection.

The differential diagnosis includes other conditions such as viral infection, tuberculosis, malign-

ant tumor, endocrine disease, amyloidosis, and connective tissue disease. Detection of ANA, anti ds-DNA antibody, and anti SS-A antibody on the eighth day of admission confirmed the diagnosis of SLE.

Of the ARA criteria for diagnosis of SLE, ANA titer is very sensitive but is non-specific for the diagnosis of lupus serositis. The SLE latex agglutination slide test and cytologic LE cell examination are complementary tools that can aid in the differential diagnosis, but are not included in the ARA criteria for SLE.⁷⁾ In the present case, the lupus erythematosus cell test was negative in both peripheral blood and pericardial effusion, although ANA was positive in both. Unfortunately, no SLE latex agglutination slide test was performed. Immunological analysis of pericardial fluid and peripheral blood from SLE patients showed lymphocytic populations and cytokine concentration pattern.⁸⁾

Cardiac tamponade is a conspicuously unusual event as an initial manifestation of SLE. The range of clinical manifestations and the outcome of pericardial tamponade in 395 patients with SLE included tamponade in 10 patients (2.5%), but tamponade was the initial manifestation of SLE in only 4 patients (1%).³⁾ In one patient, the tamponade was fatal, and 2 other patients had recurrent effusion and pericardial thickening.

Other cases of SLE presented with cardiac tamponade as the initial manifestation.^{6,9,10)} The diagnosis was based on the presence of numerous lupus erythematosus cells in the pericardial effusion. A similar case was diagnosed by the detection of excessive ANA, and confirmed by the presence of ds-DNA antibody in serum, and low complement levels in the blood.¹¹⁾ The diagnoses were confirmed by appropriate consideration of the manifestations of connective tissue diseases.

Pericarditis associated with SLE is very responsive to prednisolone.¹²⁾ If the patient is severely ill, immunoglobulin therapy is occasionally added.¹³⁾ Despite the patient's discomfort resulting from cardiac tamponade, prednisolone can be expected to smoothly improve dyspnea or shortness of breath due to pericardial effusion. If cardiac tamponade is the first manifestation, the presence of SLE remains unknown. Consequently, pericardiocentesis or pericardial window placement is indicated to improve the patient's condition.

Generally, prednisolone improves serositis in

most patients, and recurrence is rare. One case of recurrence of a moderate sized effusion required an increase in the prednisolone dosage 2 months after pericardiocentesis among 10 patients treated in this manner.³⁾ In addition, a pericardial window was indicated in 4 of 10 patients.

In summary, cardiac tamponade due to SLE pericarditis is rarely identified immediately, primarily

because of the low incidence as the initial manifestation. In addition, cardiac effusion associated with rapid sideration usually indicates bacterial infection (neutrophil dominant). The present case report suggests that careful evaluation of serous effusion is necessary to determine the most effective course of treatment.

要 約

急性心膜炎で発症し心タンポナーデをきたした全身性エリテマトーデスの1例

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症例は23歳，女性。2004年5月半ばより持続する感冒様症状が6月某日に増悪し，呼吸困難感を伴ったため当院外来を受診した。胸部X線写真上にきんちゃく様心陰影，心エコー図上で全周性の心嚢水の貯留が認められたため，精査加療の目的で当科に入院した。第1病日，心タンポナーデに進展したため，心嚢穿刺を施行した。心嚢液は膿性黄色混濁で細胞数22,100/ μ l，好中球優位(81.5%)であったため，細菌性心外膜炎を疑いカルバペネム系抗生物質の投与を開始した。しかし，翌々日に心嚢液はほぼ穿刺前の状態に戻っていた。この間37–38℃の発熱を繰り返していた。第8病日，抗核抗体5,120倍(speckled pattern)，血清抗ds-DNA抗体27.1IU/mlで，漿膜炎(心膜炎)とリンパ球減少(450/ μ l)を伴うことから，全身性エリテマトーデスによる急性心膜炎と診断した。ステロイド30mg/day内服開始，心嚢液は速やかに消失した。全身性エリテマトーデスの急性心膜炎は初発症状となることはまれで，心タンポナーデに進展することはさらに極めてまれである。また，詳しい心嚢液所見の報告例が少ないことから，文献的考察も加えて報告した。

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