CASE REPORT

# Cardiac Tamponade as Initial Presentation in Systemic Lupus Erythematosus

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## ABSTRACT

Systemic Lupus Erythematosus (SLE) is one of the many diseases known as 'the great imitators' because it can have diverse presentations and so is misunderstood for other illnesses. This case illustrates a 19 years old girl with SLE who presented as cardiac tamponade which is a rare feature of lupus pericarditis requiring medical and surgical treatment. Even after pericardiocentesis and steroid therapy there was a re-accumulation of the pericardial fluid resulting in cardiac tamponade which led to pericardial window formation. This case draws attention to the need to consider the diagnosis of tamponade in patients with connective tissue disease and dyspnea or hemodynamic compromise. It also outlines the treatment options available so that surgical referral, if needed, can be done timely for this rare but life threatening manifestation of SLE.

**Key Words:** *Lupus erythematosus systemic. Cardiac tamponade. Pericardial window technique. Pericardiocentesis.*

## INTRODUCTION

Systemic lupus erythematosus (SLE) or simply lupus is a chronic autoimmune connective tissue disease with intense inflammatory response. It can affect all organs including the heart.[1] It is characterized by formation of immune complexes and autoantibodies. There can be various forms in which the disease can present like rash, arthritis, anemia, thrombocytopenia, fever, malaise, myalgias, serositis, nephritis, seizures, and psychosis. Lupus should be kept in mind while diagnosing any patient presenting with any of these signs and symptoms, especially in female patients between 15 and 50 years of age.[2]

Cardiac involvement in SLE can present as pericarditis, myocarditis, and endocarditis. Pericarditis is the commonest cardiac complication of SLE. But cardiac involvement is rare as initial manifestation of the disease.[3] We report a case of a south asian girl who presented with cardiac tamponade and on workup was found to have SLE.

## CASE REPORT

A 19 years old girl with no known co-morbid condition presented to the emergency department with fever, chest pain, shortness of breath and generalized body swelling for 20 days. She had low grade intermittent fever, marked alopecia, joint pains and mouth ulcers for last few months. Her past medical history also revealed amenorrhea. On examination, this was a bald female sitting propped up with severe dyspnea, oral cavity examination showed ulcers on tongue and lower lip. Jugular venous pressure was raised and blood pressure was 90/60 mmHg; pulse rate was 133 per minute with respiratory rate of 30 breaths per minute and body temperature of 38 degree Celsius. On cardiovascular examination, she had gallop and heart sounds were muffled. Respiratory examination revealed bilateral crackles upto mid zones and decreased air entry at bases in both lung fields. The neurological examination was unremarkable.

Based on the history and examination, a clinical impression of SLE was made presenting with cardiac tamponade and nephrotic syndrome. Differential diagnosis was nephrotic syndrome due to minimal change disease, tuberculosis and pericardial effusion due to malignant cause. Electrocardiogram showed sinus tachycardia but there was no electrical alternans. Her initial laboratory investigations are shown in Table I. Subsequently, the echocardiogram revealed moderate to massive pericardial effusion with early diastolic right ventricle and late diastolic right atrial collapse and significant variation in mitral and tricuspid inflow (Figure I). These echocardiographic features are specific for cardiac tamponade. Her initial basic investigations revealed nephrotic range proteinuria. Her workup for SLE was sent (anti-nuclear antibody and anti-double stranded DNA). She was started on intravenous high dose pulse steroid 1 gram/day for treatment of SLE. The indication for steroid was nephrotic range proteinuria and cardiac tamponade. She was shifted to cardiac care unit where pericardiocentesis of 700 ml of straw coloured fluid was done which significantly improved her dyspnea. However, subsequently, the next day she again became dyspneic and developed type I respiratory failure. She had re-developed cardiac tamponade. She

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**Table I:** Clinical data.

| Data | Laboratory results | Normal range |
|---|---|---|
| Blood and serologic findings | | |
| Hb, gm/dl | 13.1 | 13.7 - 16.3 |
| Wbc, $mm^3$ | 13600 | 4000 - 10000 |
| Platelets $10^3$/ $mm^3$ | 184 | 150 - 400 |
| Serum albumin gm/dl | 1.6 | 3.5 - 5.5 |
| ESR | 20 | 0 - 15 |
| CRP mg/dl | 3.5 | 0 - 1 |
| ANA | Homogenous positive | |
| Serum anti-Ds DNA IU/ml | 608 | 0 - 5.3 |
| Body fluid LDH IU/L | 902 | 100 |
| Serum LDH IU/L | 1129 | 135 - 145 |
| Sodium mmol/l | 122 | 3.5 - 5.5 |
| Potassium mmol/l | 04 | 101 - 111 |
| Chloride mmol/l | 100 | 21 - 31 |
| Bicarbonate mmol/l | 18 | 0.85 - 1.35 |
| Creatinine mg/dl | 01 | 6 - 20 |
| BUN mg /dl | 60 | 42 - 121 |
| Alkaline phosphatase IU/L | 237 | |
| Pericardial fluid analysis | | |
| Glucose mg/dl | 97 | † |
| Protein mg/dl | 2900 | † |
| RBC | occasional | † |
| WBC | 800 | † |
| Lymphocytes | 30% | † |
| Polymorphs | 70% | † |
| Kidney biopsy | | |
| Lupus type 4 nephritis | | |
| IgG | Positive | |
| IgM | Positive | |
| IgA | Positive | |
| C3 | Negative | |
| C1 | Positive | |
| Urinary protein g/L | Greater than 3.5 (+3) | |
| Spot urine protein mg/dl | 1148 | |

*WBC = White blood cells; Hb = Hemoglobin; ESR = Erythrocyte sedimentation rate; CRP = C-Reactive protein; ANA = Antinuclear antibody; RBC = Red blood cells; † Normal values for pericardial fluid no established.*

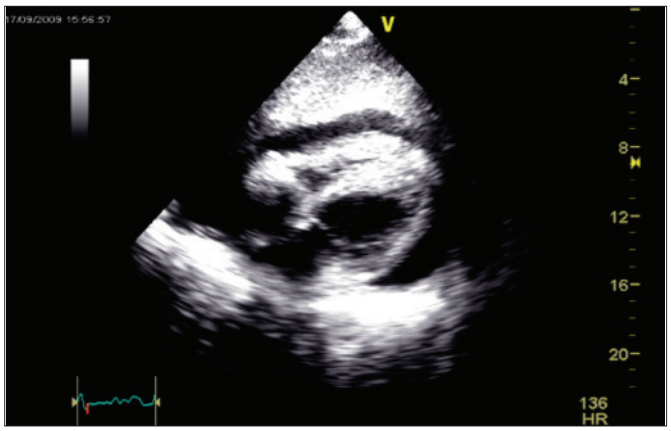


**Figure 1:** Large circumferential pericardial effusion in sub-costal view.

was intubated due to persistent hypoxia of 75% $O_2$ saturation on room air. Due to recurrence of cardiac tamponade, she was taken for surgery and a pericardial window was formed by the cardiothoracic surgeon. A chest tube was also inserted on the left side. Subsequently, she improved on steroids and was extubated the next day and shifted out of cardiac care unit with drains in place.

She later had a renal biopsy which revealed diffuse proliferative glomerulonephritis (features compatible with WHO class four lupus nephritis). She was shifted to oral steroids and azathioprine was added as steroid sparing agent. Her pleural and pericardial tubes were later removed and she was discharged home.

## DISCUSSION

We report here a case of SLE with initial presentation of cardiac tamponade which required pericardial window apart from medical treatment. The unique thing was that this was a very aggressive presentation of SLE which required a surgery for treatment. Connective tissue diseases usually do not have an abrupt life threatening manifestation.

In a retrospective study which reviewed cases between 1985 - 2006 found that cardiac tamponade occurred in only 9 out of 41 SLE patients who presented with pericardial effusion. All of them were women and one was Asian. Five out of them required pericardial window inspite of being treated with high dose corticosteroids.[4] One case of a young girl with SLE presenting as cardiac tamponade has also been reported from India.[5] This is the first case being reported from Pakistan.

Cardiac tamponade occurs when a critical amount of fluid accumulates in the pericardium resulting in decreased blood flow to the ventricles. Typical symptoms and signs include dyspnea, orthopnea, chest pain, pulsus paradoxsus and hypotension. Tamponade was seen in < 1% patients of the combined series of more than 1300 patients.[6] It was also found that the likely predictors of tamponade in SLE patients who present with pericarditis and pericardial effusions are low serum C4 levels, female gender, concurrent renal disease, hemolytic anemia and pleurisy.[4]

Frequency of pericardial involvement in different connective tissue diseases varies but is highest in systemic sclerosis and SLE being 60% and 44% respectively, rheumatoid arthritis and mixed connective tissue disease (30% and 24%), polymyositis, dermato-myositis and rheumatoid arthritis (24% and 11%).[7] It can produce complications like cardiac tamponade, purulent and constrictive pericarditis.

Pericardiocentesis is a good option for large effusions, but recurrence is common and in that case a less invasive approach called pericardial window can be applied in which a fistula is made connecting pericardial space with pleural space advantage of which can be possible tissue biopsy of the pericardium and prevention of recurrences. Pericardiectomy is also an option if there

is future accumulation of pericardial fluid after pericardiocentesis and steroid therapy and is also helpful in preventing future symptoms of constrictive pericarditis.[8]

This patient underwent pericardial window formation within 48 hours of presentation. A young male patient in whom cardiac tamponade secondary to loculated pericardial effusion was the presenting symptom of SLE had an emergency pericardiocentesis and pericardiectomy at the same session to prevent future complications. He had an uneventful postoperative course.[9]

NSAIDS and corticosteroids are effective in this rare hemodynamic effect of pericarditis in patients of multiple connective tissue disorder. This might also be a reason for low incidence of tamponade despite high frequency of pericardial disease on autopsy and echocardiographic findings. Pericardiocentesis is reserved for persistent large pericardial effusion and potentially life threatening tamponade. Non-responders require pericardial window.[10] Cardiac tamponade in systemic lupus erythematosus should be treated with high dose steroids and pericardiocentesis but when pericardiocentesis is impossible to attempt, intravenous immunoglobulins can also be used as another treatment option.

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