

## Case Report

# A 16-Year-Old Girl with Systemic Lupus Erythematosus (SLE) with Septicaemia as Initial Presentation

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### Abstract

*Sepsis is a life-threatening condition caused by systemic host response to infection. Sepsis or septic shock is considered to be rare as initial presentation of autoimmune disease like systemic lupus erythematosus (SLE). SLE is a multisystem disorder which usually occurs in adolescent girls. So when an adolescent girl presents with septicaemia and multi-organ dysfunction, an autoimmune disease like SLE should be excluded. Here we report a 16-year-old girl initially presenting with septic shock along with multiorgan dysfunction. The patient was thoroughly evaluated and shock was managed effectively according to protocol. Later on she was diagnosed as a case of systemic lupus erythematosus (SLE).*

**Key words:** Systemic lupus erythematosus (SLE); Septicaemia; Multiorgan dysfunction

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## Introduction

Septicaemia can be defined as systemic inflammatory response to infection and when it is complicated with organ dysfunction it is termed as severe sepsis.<sup>1</sup> On the other hand, septic shock is described as sepsis complicated with hypotension that is refractory to fluid resuscitation.<sup>2</sup> Sepsis is the common cause of morbidity and mortality in patient with systemic lupus erythematosus (SLE) due to the altered immunity, increased dose of steroid use and hypocomplementaemia.<sup>3,4</sup> It has been shown that incidence of septicaemia is ten times more common in SLE patients than in non-SLE group.<sup>6</sup> But features of infection like high fever, loose motion or cough with respiratory distress are not usual at the onset of pediatric SLE. So evaluation of SLE should be considered in the patient with fever of unknown origin with different organ involvement and raised ESR.<sup>5</sup> As the prognosis of SLE patient with septicaemia is guarded, early diagnosis, effective management and meticulous

follow-up are needed to improve the outcome of this condition.<sup>6</sup>

## Case report

A 16-year-old immunized girl was admitted in the Department of Paediatrics, Bangabandhu Sheikh Mujib Medical University (BSMMU) with the complaints of high grade continued fever and non-productive cough for seven days. She also had history of non-bloody watery diarrhoea for several times along with diffuse abdominal pain for last two days and anuria for last 24 hours. For the last 12 hours she developed severe respiratory distress along with restlessness. She had no history of convulsion, unconsciousness, previous urinary complaints or contact with tuberculous patient. She was diagnosed as a case of nonspecific musculoskeletal pain syndrome two years back and got treatment, but she did not continue follow-up. On examination she was restless, febrile and mildly

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pale. Features of shock were present in the form of cold clammy skin, tachycardia with low volume pulse (140 beats/minute), tachypnoea (respiratory rate 38 breaths/minute), prolonged capillary refill time (CRT 3.5 seconds), low oxygen saturation (SPO<sub>2</sub> was 88% in room air) and low blood pressure (70/40 mm Hg). Her bed side ESR was 120 mm in 1<sup>st</sup> hour and bed side urine albumin was nil. Capillary blood glucose was 4.5 mmol/L. There was diffuse abdominal tenderness with normal bowel sound, non-tender hepatomegaly and bilateral pleural effusion. She was diagnosed as a case of septicaemia with shock with multiorgan dysfunction.

Her immediate investigation reports revealed anaemia with neutrophilic leukocytosis with raised ESR (haemoglobin 10.9 gm/dL, WBC 22000/cmm, platelet 450,000/cmm, ESR 120 mm in 1<sup>st</sup> hour), proteinuria (++) and mild respiratory alkalosis (pH 7.47, partial pressure of CO<sub>2</sub> 33 mm Hg, partial pressure of O<sub>2</sub> 81 mm Hg, bicarbonate 23.2

mmol/L). Chest radiography showed right sided pleural effusion with opacities in both lung fields (Fig 1). There was raised lactate dehydrogenase (521 U/L), serum ferritin (>40000 mg/mL), fibrin degradation product (97.7 mg/dL) and prothrombin time (20.7 second), INR 1.73, normal serum creatinine (0.53 mg/dL), electrolytes (Na<sup>+</sup> 137 mmol/L, K<sup>+</sup> 3 mmol/L, Cl<sup>-</sup> 107 mmol/L, TCO<sub>2</sub> 22 mmol/L), activated partial thromboplastin time (27.6 second) and serum triglyceride level (112 mg/dL). Antinuclear antibody was found negative. Her blood and urine culture revealed no growth. Initially she was immediately managed with normal saline bolus (20 mL/kg) followed by maintenance with 5% dextrose in cholera saline along with oxygen inhalation (Fig 2). Injection ceftriaxone and amikacin were started along with omeprazole, vitamin K and fresh frozen plasma. But the patient had persistent respiratory distress, high fever along with more severe abdominal pain and bipedal oedema.



Fig 1. Right sided pleural effusion with opacity in hilar region



Fig 2. Immediate resuscitation

Radiography of abdomen was normal. Antibiotic was changed to injection meropenem, vancomycin and metronidazole. For persistent hypotension injection dopamine followed by noradrenalin was started according to protocol. Also injection hydrocortisone was added to improve the blood pressure. Vitals were closely monitored and continuous catheterisation was ensured. With these measures blood pressure improved though respiratory distress and abdominal pain persisted. So metronidazole was omitted and ciprofloxacin was added. After adding ciprofloxacin abdominal pain subsided.

On the 6<sup>th</sup> day she developed recurrent apnoea. There was no heart beat and respiratory effort. Cardiopulmonary resuscitation along with bag mask ventilation was given. Around five minutes after resuscitation patient improved but there was bradyarrhythmia (heart rate 48 beats/min, irregular). So noradrenalin was stopped gradually and some investigations were done. This time investigation reports revealed anaemia, neutrophilic leukocytosis and raised ESR (haemoglobin 9.6 gm/dL, WBC 35000/cumm, N 94%, L 3%) with hypoalbuminaemia (serum albumin 1.4 gm/dL). Electrolytes, calcium and random blood glucose were normal (S electrolytes: Na<sup>+</sup> 144 mmol/L, K<sup>+</sup> 4.1 mmol/L, Cl<sup>-</sup> 105 mmol/L; S. Ca<sup>++</sup> 1.68 mmol/L; RBS 7.6 mmol/L). ECG was normal. Anti-ds DNA was positive (125 U/mL). Urinary total protein was also significantly raised (UTP 0.74 gm/day) and Coomb's test was negative. According to the clinical picture and laboratory profile she was diagnosed as a case of systemic lupus erythematosus with septicemia. So tablet hydroxychloroquine along with oral prednisolone were started. With these medications she gradually improved. Vital signs and oxygen saturation were normal. So she was shifted to paediatric rheumatology unit for further evaluation.

## Discussion

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease causing multisystem involvement.<sup>8</sup> SLE patients are highly susceptible to infection due to the combined effect of immunosuppressive therapy as well as disease-induced abnormalities of the immune system.<sup>8</sup> Hospitalisation rates of SLE patients due to sepsis has been

substantially increased from 1996 to 2011.<sup>9</sup> Also in-hospital mortality is higher in SLE patients, especially those with opportunistic infections and pneumonia or sepsis requiring mechanical ventilation.<sup>9</sup> Although pneumonia, urinary tract and skin infection are commonly reported in SLE patients, bacteraemia or sepsis complicated with organ failure is the leading cause of hospital mortality.<sup>10,11</sup>

In a previous study sepsis-induced mortality was observed in 30% patients who had multiorgan dysfunction syndrome and 60–80% patients who initially presented with septic shock.<sup>12</sup> The success of the treatment in sepsis depends on early diagnosis, immediately starting appropriate antibiotic along with supportive management and diagnosis as well as treatment of the underlying aetiology.<sup>13</sup>

Our patient initially presented with features of sepsis along with shock and multiorgan dysfunction. Her investigations were also suggestive of septicemia though blood and urine culture revealed no growth of any organisms. As she was an adolescent girl with multisystem involvement, antinuclear antibody (ANA) was done initially to exclude SLE, which was found negative.

We also kept macrophage activation syndrome (MAS) as our differential, which is a common life-threatening complication in several autoimmune diseases including SLE.<sup>20</sup> The laboratory profile of MAS should include cytopenia with sudden drop of ESR in complete blood count, hypertriglyceridaemia and raised serum ferritin (>5000–10,000 ng/mL).<sup>21,22</sup> Though our patient had high serum ferritin level (>40,000 ng/mL), presence of high ESR and normal triglyceride exclude macrophage activation syndrome in our patient.

The main target of treatment for septic shock is to maintain blood pressure and also tissue perfusion.<sup>14</sup> So after fluid resuscitation vasoactive drugs are needed to improve the blood pressure and pulse volume.<sup>15</sup> Antibiotics are given empirically until identification of causative organism.<sup>14</sup> Corticosteroid is another important agent in sepsis treatment which may reduce the death risk in these patients.<sup>16</sup>

After admission we initially managed our patient

with normal saline bolus followed by dopamine infusion. We also started injection ceftriaxone and amikacin empirically which were changed to meropenem and vancomycin due to persistent high fever. Steroid was also added. Later on ciprofloxacin was added as gastroenteritis was suspected to cause persistent abdominal pain. When blood pressure was not improved after adding dopamine and steroid, norepinephrine was added. Norepinephrine, a potent alpha adrenergic agent, is an accepted treatment for dopamine unresponsive septic shock.<sup>17</sup> Previously it was thought that epinephrine affects the cardiac contractility and may cause cardiac arrest which was not a usual side effect of norepinephrine.<sup>18</sup> But later on a study reported that cardiac arrest was more in norepinephrine group than in epinephrine group when these catecholamines were used to treat septic shock.<sup>19</sup>

In our case when blood pressure was not improved with dopamine infusion, we started norepinephrine. But after adding norepinephrine patient developed recurrent apnoea along with cardiac arrest. So after cardiopulmonary resuscitation we gradually stopped the norepinephrine infusion and the patient settled.

After stabilisation, we re-evaluated our patient for SLE as she was an adolescent girl, had previous history of recurrent arthralgia and presented with multisystem involvement including hepatomegaly, pleural effusion and proteinuria. We did anti ds-DNA which was positive and also did 24-hour urinary total protein (UTP) which was significantly raised. So finally patient was diagnosed as a case of systemic lupus erythematosus (SLE).

Separating the acute episode of SLE from sepsis on emergency ground is considered to be the most challenging situation.<sup>23</sup> In emergency situation when an adolescent girl presents with acute life-threatening condition with multisystem involvement, a suspicion of autoimmune disease particularly SLE should be considered.<sup>24</sup>

Along with prolonged fever and constitutional symptoms, the mucocutaneous (malar rash, alopecia, oral ulcer, photosensitivity) and musculoskeletal involvement (arthritis or arthralgia) are the usual presentations of SLE.<sup>25</sup> Initial presentation with sepsis along with shock and multiorgan dysfunction

in SLE is rare.<sup>26</sup>

The laboratory evidence of SLE may include low hemoglobin with raised ESR, proteinuria, features of serositis (pleural or pericardial effusion or ascites) and a positive ANA. Though ANA is highly sensitive for SLE, it is less specific.<sup>27</sup> Testing for antibody to anti-ds DNA and antibody to Sm nuclear antigen (Anti-Sm) may be helpful in patients whose clinical and laboratory features are suggestive of SLE but ANA is found negative.<sup>28</sup> Initially our patient had raised ESR, proteinuria and pleural effusion with negative ANA. Though ANA was negative, after managing septic shock we re-evaluated our patient for SLE by doing anti-ds DNA and UTP, both of which were found raised.

Once SLE is diagnosed the mainstay of treatment is hydroxychloroquine along with corticosteroid in the form of oral prednisolone or intravenous methyprednisolone.<sup>25</sup> But prednisolone may flare up the infection. So after controlling infection we started hydroxychloroquine and also switched hydrocortisone to oral prednisolone. Prognosis of these patients depends on treatment response. Good response to treatment may improve outcome of this disease.<sup>5</sup>

## Conclusion

SLE with septicaemia and multiorgan dysfunction is a life-threatening condition with high mortality rate. Rapid diagnosis, early initiation of treatment and regular monitoring of clinical and laboratory profile are needed for proper management of these patients. Above all in case of adolescent girls presenting with septicaemia and multiorgan dysfunction, SLE in background should be in mind.

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