Case report

Two adult cases of sepsis-associated purpura fulminans

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(Received: December 8, 2014; Accepted: March 30, 2015)

Summary

Two uncommon cases of sepsis-associated Purpura fulminans (PF) caused by urinary tract infection are herein reported. The first case involved an 81-year-old man, and the second an 80-year-old woman; both developed septicemia and disseminated intravascular coagulation (DIC). After admission to the intensive care unit (ICU), progressive purpuric skin necrosis became evident on the fingers and toes. Blood and urine cultures were positive for both Escherichia coli and Enterococcus faecalis in the first case and for Escherichia coli in the second. The patients were discharged from the ICU on day 17 and 36, respectively. All areas of necrosis subsequently separated from the underlying tissue. Because the mortality rate associated with PF is very high, priority must be given to general and symptomatic supportive therapy in patients with sepsis-associated PF.

Introduction

Purpura fulminans (PF) is a rare disease characterized by the rapidly progressive development of purple skin lesions due to disseminated intravascular coagulation syndrome (DIC) and microvascular thrombosis. PF is associated with congenital or acquired deficiency of coagulation factors such as protein C or protein S. PF with infectious disease in particular is called acute infectious PF (AIPF), which is a rare complication of severe sepsis. We describe two uncommon cases of sepsis-associated PF secondary to urinary tract infection.

Case presentation

An 81-year-old man was brought by ambulance to our emergency medical care center and admitted to the intensive care unit (ICU) because of septic shock and DIC after a 3-day period of pyrexia. His medical history included diabetes mellitus, prostatic carcinoma, and left urinary tract lithiasis. Blood examination showed severe leukocytopenia and thrombocytopenia, an increased inflammatory reaction, and acute renal failure. His Japanese Association for Acute Medicine (JAAM) DIC score was 8 points; a score of >5 points fulfills the revised JAAM criterion for sepsis-induced DIC. Arterial blood gas analysis showed hypoxemia and metabolic acidosis (Table 1). Whole-body computed tomography showed a left ureteral stone and hydronephrosis with increased density around the left kidney. The patient’s sequential organ failure assessment (SOFA) score was 12 points, and severe sepsis was diagnosed. After tracheal intubation under general anesthesia to allow for respiratory management, the patient was started on a continuous infusion of noradrenaline and gabexate.

Key words: sepsis-associated purpura fulminans, septicemia, septic shock, disseminated intravascular coagulation, urinary tract infection
**Table 1** Laboratory data

<table>
<thead>
<tr>
<th></th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 1</th>
<th>Case 2</th>
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<tbody>
<tr>
<td>RBC ($\times 10^{12} / \mu L$)</td>
<td>403</td>
<td>401</td>
<td>340</td>
<td>198</td>
<td>Blood gas analysis*</td>
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<tr>
<td>Hb (g/dL)</td>
<td>12.8</td>
<td>11.5</td>
<td>0.8</td>
<td>0.4</td>
<td>pH</td>
<td>7.459</td>
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<tr>
<td>Hct (%)</td>
<td>37.8</td>
<td>35.4</td>
<td>0.4</td>
<td>0.1</td>
<td>$\text{PaCO}_2$ (mmHg)</td>
<td>20.8</td>
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<tr>
<td>WBC ($\times 10^{9} / \mu L$)</td>
<td>0.4</td>
<td>15.0</td>
<td>47.2</td>
<td>60.3</td>
<td>$\text{PaO}_2$ (mmHg)</td>
<td>57.6</td>
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<tr>
<td>Platelets ($\times 10^{9} / \mu L$)</td>
<td>3.3</td>
<td>8.4</td>
<td>2.2</td>
<td>5.3</td>
<td>HCO$_3$ (mmol/L)</td>
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<td>APTT (sec)</td>
<td>40.5</td>
<td>44.1</td>
<td>132</td>
<td>137</td>
<td>Base excess (mmol/L)</td>
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<td>PT-INR</td>
<td>1.38</td>
<td>1.59</td>
<td>4.4</td>
<td>4.0</td>
<td>Glucose (mg/dL)</td>
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<td>Fibrinogen (mg/dL)</td>
<td>762</td>
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<td>102</td>
<td>104</td>
<td>Lactate (mmol/L)</td>
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<td>58.0</td>
<td>57.0</td>
<td>38.1</td>
<td>34.1</td>
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<td>FDP ($\mu g$/mL)</td>
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<td>138.2</td>
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<td>AST (IU/L)</td>
<td>28</td>
<td>88</td>
<td>1988</td>
<td>45</td>
<td>SOFA score</td>
<td>12</td>
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<tr>
<td>ALT (IU/L)</td>
<td>14</td>
<td>28</td>
<td>$&lt;5.00$</td>
<td>$&lt;5.00$</td>
<td>JAAM DIC score</td>
<td>8</td>
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<td>LDH (IU/L)</td>
<td>226</td>
<td>437</td>
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</tbody>
</table>

*Case 1: arterial blood and room air; Case 2: venous blood and 100% nasal oxygen at 2 L/min

RBC, red blood cells; Hb, hemoglobin; Hct, hematocrit; WBC, white blood cells; APTT, activated partial thromboplastin time; PT-INR, prothrombin time-international normalized ratio; AT-III, antithrombin-III; FDP, fibrin degradation products; AST, aspartate aminotransferase; ALT, alanine aminotransferase; LDH, lactate dehydrogenase; ALP, alkaline phosphatase; T-bilirubin, total bilirubin; D-bilirubin, direct bilirubin; BUN, blood urea nitrogen; Na, sodium; K, potassium; Cl, chloride; CRP, C-reactive protein; CPK, creatine phosphokinase; B-D glucan, beta-D-glucan; $\text{PaCO}_2$, partial pressure of carbon dioxide; $\text{PaO}_2$, partial pressure of oxygen; HCO$_3$, bicarbonate; SOFA score, Sequential Organ Failure Assessment score; JAAM DIC score, revised Japanese Association for Acute Medicine for sepsis-induced disseminated intravascular coagulation score

![1A](image1.png) ![1B](image2.png) ![1C](image3.png) ![1D](image4.png)

**Figure 1** The photographs of Case 1

The photographs show the patient’s hands and feet on the 10th day after admission (1A: left hand, 1B: right hand, 1C: right foot, 1D: left foot).

Mesylate as well as boluses of antithrombin-III, $\gamma$-globulin, and meropenem. A urologist placed a stent in the left ureter, and endotoxin adsorption was started. Fresh frozen plasma, platelets, and red blood cell concentrate were administered. *Escherichia coli* and *Enterococcus faecalis* were detected in urine and blood cultures. Purple spots appeared on the peripheral limbs during the following 10 hours, then gradually deteriorated. The DIC resolved on the 10th day after admission, and the patient was given warfarin, which had been prescribed before admission. The limb necrosis became localized to the tips of the toes and fingers, then progressed to dry necrosis (Fig. 1). The patient was discharged from the ICU on day 17.

A healthy 80-year-old woman was transferred by ambulance from a local hospital to our emergency medical care center and admitted to the ICU. She was suspected of having septicemia and DIC (Table 1). Her JAAM DIC score was 6 points. Computed tomography revealed bilateral ureteral stones and hydronephrosis. The enlarged left renal pelvis was considered to be an acute change because of the high density around the kidney. The patient’s SOFA score was 8 points, and severe sepsis was diagnosed. After placement of a stent into the left ureter, continuous gabexate mesylate and boluses of meropenem, antithrombin-III, and $\gamma$-globulin were administered. Endotoxin adsorption was performed, and continuous hemodiafiltration was started.
for persistent oliguria. Platelets and red blood cell concentrate were administered for progressive thrombocytopenia and anemia. Because her respiratory function deteriorated, tracheal intubation was performed and respiratory management was begun. *Escherichia coli* was detected in both urine and blood cultures on day 4. Paleness and purple spots appeared on the peripheral parts of the lower limbs 15 hours after admission and on the upper limbs 33 hours after admission, then gradually deteriorated. The DIC resolved on day 7, and prostaglandin E₁ therapy was started. The limb necrosis decreased and became localized to the tips of the toes and fingers, finally progressing to dry necrosis (Fig. 2). Although the patient was extubated on day 11, reintubation was needed to maintain the airway due to vocal cord paralysis. A tracheotomy was subsequently performed, and she was taken off the respirator. Following an oliguric phase, diuresis ensued and her renal insufficiency improved. She was discharged from the ICU on day 36.

Discussion

PF is a rare disease characterized by rapidly progressive development of purple skin lesions secondary to DIC and microvascular thrombosis. It is defined as simultaneous ischemic necrosis of more than two affected distal limbs without obstruction of proximal arteries.² Because proximal arterial obstruction was not present in either affected limb, our patients’ conditions fit the definition of PF.

PF includes congenital (inherited) PF, which is a hereditary disease; idiopathic (post-infectious) PF, which develops after a preceding infection such as chickenpox or scarlet fever; and AIPF, which is caused by bacterial infection.¹⁻³ Previous reports on AIPF have shown that the main pathogens are *Neisseria meningitidis*, *Streptococcus pneumoniae*, Group A and B streptococci, *Haemophilus influenzae*, and *Staphylococcus aureus⁴⁻⁵*, however, *Escherichia coli* and *Enterococcus faecalis* are rare.⁶

The mortality rate of PF is >50%, and the main causes of death are multiple organ failure and DIC with adrenal bleeding. Patients who survive often require amputation of dry gangrene that progresses from purpura on the distal limbs, which has a poor functional prognosis.⁷ Many case reports and small series on the treatment of PF have been published, but no large studies have attempted to identify a definitive cure. Therefore, priority must be given to general and symptomatic supportive therapy in patients with severe sepsis-associated AIPF. All areas of necrosis eventually separated from the underlying tissue in both of the herein-described cases, but prostaglandin E₁ therapy helped to considerably reduce the necrotic area in the

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*Figure 2* The photographs of Case 2

The photograph on the right side shows the patient’s right hand on the 7th day after admission (2A). The photographs on the left side show the same hand after prostaglandin E₁ therapy (2B: Day17, 2C: Day29).
second case.

Conflict of interests

The authors declare no relevant financial or material conflicts of interest.

References