

Acute Infectious Purpura Fulminans Complicated with Multiple Erosions

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Abstract An 80-year-old man was extremely skinny and developed a left leg phlegmon. He was admitted and underwent an infusion of cefazolin. On the third day, he suddenly collapsed, and advanced cardiac life support resulted in a return of his circulation. He showed vasopressor-resistant hypotension and therefore received an infusion of steroids and multiple supportive therapies. A blood culture later showed *Pseudomonas aeruginosa*. While his blood pressure increased, he showed peripheral and labile cyanotic changes, even though his peripheral arteries were palpable. On the third day after the collapse, he showed blisters at purpura sites and labile, resulting in the formation of erosions. On the fourth day, massive effusion was drained from the erosive lesions. He temporarily showed a return of consciousness, but his extremities became necrotic, and he ultimately died due to a secondary infection at the erosive lesions. The present fatal case of acute infectious purpura fulminans might have been induced by a *P. aeruginosa* infection due to complications with multiple blisters, similar to Stevens-Johnson syndrome/toxic epidermal necrolysis. Early aggressive surgical amputation or debridement might be required to obtain a survival outcome when encountering such cases.

Keywords: *purpura fulminans, Pseudomonas aeruginosa, toxic epidermal necrolysis*

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1. Introduction

Acute infectious purpura fulminans (AIPF) is a rare syndrome of hemorrhagic infarction of the skin, manifesting as a symmetric peripheral gangrene resulting in extremity loss, disseminated intravascular coagulation and multiple organ failure induced by infection. [1] The two predominantly identified microorganisms are *Neisseria meningitidis* and *Streptococcus pneumoniae*. [2,3] AIPF has a high mortality rate and miserable functional outcome due to amputation, even when patients obtain a survival outcome. [2]

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are life-threatening diseases characterized by detachment of the epidermis and mucous membrane, occurring mainly due to drugs. SJS/TEN are considered on the same spectrum of diseases, only demonstrating different severities. [4] AIPF may be complicated with extensive blisters, similar to those seen in cases of SJS/TEN. [5,6]

We herein report a rare case of AIPF with blisters resembling SJS/TEN induced by *Pseudomonas (P)aeruginosa*.

2. Case Presentation

An 80-year-old man who was extremely skinny due to post-operative esophageal gastrointestinal stromal tumor

and had leg edema due to deep venous thrombosis presented with left leg phlegmone. He was admitted to the Dermatology Ward and received infusion of cefazolin. However, his systolic blood pressure decreased to 60 mmHg which thus made it necessary to place the patient in the shock position. The next morning, he suddenly collapsed, and a code blue alert was sounded. Tracheal intubation, infusion of 1 mg of adrenaline and 1 cycle of chest compression resulted in a return of circulation being obtained. He underwent a blood examination and enhanced whole-body computed tomography (CT) and was transferred to the intensive-care unit. The results of blood examinations showed anemia, thrombocytopenia, hypoproteinemia, hyperglycemia, rhabdomyolysis, renal failure and coagulopathy (Table 1). CT showed only bilateral pleural effusion and atelectasis. An electrocardiogram showed sinus tachycardia without ST changes. He showed vasopressor-resistant hypotension and therefore received infusion of steroid in addition to noradrenaline and vasopressin. He also received infusion of meropenem, vancomycin and gamma globulin instead of cefazolin and was transfused with red blood cells, platelets and fresh-frozen plasma. Culture of blood later showed *P. aeruginosa*. His blood pressure increased under these treatments. However, he showed peripheral and labile cyanotic changes, even though his peripheral arteries were palpable. On the third day after the collapse,

he showed blisters at purpura sites and labile, resulting in the formation of erosions. He also showed black gastric fluid from the nasogastric tube and black diarrhea, suggestion hemorrhaging from the bowels. On the fourth day, massive effusion (over 3 L) was drained from the erosive lesions. He temporarily showed a return of consciousness, but his extremities became necrotic (Figure 1), and he ultimately died on day 21 due to secondary infection at the erosive lesions. Permission to perform an autopsy was not obtained from his family.

3. Discussion

We performed a Medline search to identify any related articles using the key words “purpura fulminans” and “*P. aeruginosa*”. As a result, we found two reports: one by Lerolle et al. and one by Aroor et al. [7,8] Lerolle et al. reported the results of fluorescence confocal microscopy, which showed *P. aeruginosa* in the dermal vessels of a deceased patient with purpura fulminans. [7] Microorganisms

were typically identified inside the capillaries in foci with multiple bacteria in the purpuric skin. [7] In their report, the clinical course was not described except for the patient’s ultimately death. Aroor et al. reported an eight-year-old boy who presented with a fever and purpuric rash involving all four limbs. [8] Their patient eventually developed gangrene of all digits and the pinna, and the condition deteriorated progressively until he was discharged against medical advice in a moribund state. In that case, the blood culture grew *P.*, but whether or not it was *P. aeruginosa* was unknown. Accordingly, the present case might be the second or third case of AIPF induced by *P. aeruginosa*.

Risk factors for AIPF in the present case might have included malnutrition, old age and a history of skin disease. [9,10] As the patient had vasopressor-resistant hypotension requiring steroids, he might have had adrenal insufficiency in a critical state or Waterhouse-Friderichsen syndrome. [11,12] As he had received infusion of cefazolin before his collapse, the potential involvement of some undetected causative bacteria cannot be ruled out.

Table 1. Blood biochemistry on arrival

white blood cell 9,700/ μ l	hemoglobin 7.8 g/dl	platelet 7.5×10^4 / μ l
total protein 3.6g/dl	albumin 1.7 g/dl	total bilirubin 2.1 mg/dl
glucose 232 mg/dl	amylase 31 IU/l	creatinine kinase 283 IU/l
sodium 139 mEq/l	potassium 3.7 mEq/l	chloride 113 mEq/l
aspartate aminotransferase 75 IU/l	alanine aminotransferase 27 IU/l	
blood urea nitrogen 29.5 mg/dl	creatinine 1.10 mg/dl	lactate dehydrogenase 323 IU/l
C-reactive protein 16.4 mg/dl	prothrombin time 50.1 (11.6) sec	
activated partial thromboplastin time 79.8 (27.2) sec		
fibrinogen 185 mg/dl	fibrinogen degradation products 9.2 μ g/ml	



Figure 1. Lower extremities of the present case. The bilateral legs showed purpura and necrosis with multiple skin erosions

The present case developed multiple erosions at purpura lesions, resulting in massive effusion resembling an extended burn, and secondary infection at these lesions led to his death. Patients with atopic dermatitis whose skin barrier is weak to bacterial infection might become complicated with staphylococcal scalded skin syndrome (SSSS)-like blister lesions induced by *P. aeruginosa* infection. [13] As the present case had a history of skin lesions and complication with *P. aeruginosa* infection, his multiple blister lesions might have been complicated with a *P. aeruginosa* infection.

Mazzone et al. reported that the survival in cases of AIPF was not dependent on surgery, suggesting that surgery does not play a key role in the early phase of the disease and that debridement should be postponed until clear demarcation has been established. [14] However, all patients with AIPF complicated with multiple erosions resembling SJS/TEN who underwent supportive therapy without early surgical operation died, similar to the present case. [5,6,15] Accordingly, early aggressive surgical amputation or debridement may be required to obtain a survival outcome, even in patients with serious sequelae.

4. Conclusion

The present fatal case of AIPF might have been induced by *P. aeruginosa* infection and complicated with SJS/TEN. Early aggressive surgical amputation or debridement may be required to obtain a survival outcome in such combination cases.

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Statement of Competing Interests

We do not have no competing interests.

List of Abbreviations

AIPF: Acute infectious purpura fulminans
 CT: computed tomography
 SJS: Stevens-Johnson syndrome
 SSSS: staphylococcal scalded skin syndrome

TEN: toxic epidermal necrolysis

P: *pseudomonas*

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