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Ecthyma gangrenosum associated with aplastic anemia

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Ecthyma gangrenosum is a characteristic skin lesion of systemic infection due to Pseudomonas aeruginosa. It has a high incidence in patients with chronic disease and impaired defense mechanisms. Early diagnosis and appropriate systemic antibiotic therapy is crucial since its mortality rate is very high. We report a case of ecthyma gangrenosum in aplastic anemia.

Key Words : *Ecthyma gangrenosum, Pseudomonas aeruginosa, Aplastic anemia*

INTRODUCTION

Ecthyma gangrenosum is a characteristic skin lesion of a potentially life-threatening systemic infection due to *Pseudomonas aeruginosa*. This lesion was first described by Barker(1897) and subsequently noted by other authors to be pathognomonic of Pseudomonas septicemia(Hitschmann and Kreibich, 1897). Sepsis due to this organism arises particularly in critically ill patients who are immunologically impaired. The skin manifestations of ecthyma gangrenosum begin as a painless round erythematous macule which becomes indurated, bullous or pustular. It develops into a round, indurated, painless area with central necrotic black or gray-black eschar and a surrounding erythematous halo(Greene et al., 1984a). We describe a patient with aplastic anemia who developed striking skin lesions associated with Pseudomonas infection.

CASE REPORT

A 36-year-old female with aplastic anemia was admitted because of sudden onset of right lower quadrant pain, fever, chills, general weakness, nausea, vomiting, and diarrhea for two days. The diagnosis of aplastic anemia was made ten years previously on the basis of peripheral blood count showing pancytopenia and a bone marrow biopsy revealing hypocellular marrow. Immunotherapy was given five years later. It consisted of anti-thymocyte globulin 300mg/day for 5 days, cyclosporine A 600mg/day for two weeks, solumedrol 300mg IV/day for 7 days, oxymetholone 50mg/day for three weeks, pyridoxine 300mg/day for three weeks, and prednisolone 30mg/day for three weeks concurrently. There were no complications and the patient received intermittent RBC transfusions on an outpatient basis. On the first hospital day the patient experienced an exacerbation of right lower quadrant pain.

The patient was alert with an acutely ill looking appearance. Blood pressure was 90/70mm Hg ; pulse rate, 116 per minute ; temperature 37.8℃ orally. Pertinent physical findings were direct and indirect tenderness of the right lower quadrant and decreased bowel sounds on auscultation. Hematocrit and hemoglobin levels were 29 % and 9.8g/dl. White blood cell count was 390/microliter and the platelet count was 81,000/microliter. A laparoscopic appendectomy was performed under the impression of acute appendicitis. The appendix showed signs of

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severity depend on the patient's immunologic status. A healthy person may have a relatively minor infection such as intertrigo, green nail syndrome, or folliculitis. In immunocompromised patients, Pseudomonas species are a common cause of septicemia(Sevinsky et al., 1993).

Ecthyma gangrenosum is one of the characteristic cutaneous manifestations in immunocompromised patients, particulary those with underlying malnutrition, malignant disease, or severe hematologic disease and those who have undergone antibiotic or immunosuppressive therapy(Huminer et al., 1987).

Pseudomonas aeruginosa is a saprophytic intestinal gram-negative aerobic rod with a predilection for human environments(Sevinsky et al., 1993). Barker(1897) first recognized that in sepsis due to *Pseudomoas aeruginosa,* characteristic skin lesions arose and developed from erythematous macules or vesicles into necrotic ulcers after systematically reviewing bacteriological findings in 800 consecutive autopsies at the Johns Hopkins Hospital. In the same year Hitschmann and Kreibich(1897) gave the name "ecthyma gangrenosum" to the typical skin lesions associated with *Pseudomoas aeruginosa.* Kerby(1947) reviewed eighty-three previously reported cases of septicemia due to *Pseudomoas aeruginosa.* In the 1950s and the 1960s, it became apparent that a significant number of patients with cancer died from *Pseudomoas aeruginosa* septicemia. This organism was responsible for 25 % of the fatal infections in patients with malignant diseases(Hersh et al., 1965).

The clinical skin manifestations of ecthyma gangrenosum begin as a painless round erythematous macule which becomes indurated, bullous or pustular. It develops into a round, indurated, painless area with central necrotic black or gray-black eschar and surrounding erythematous halo(Greene et al., 1984a). These lesions occur more frequently in the anogenital, axillary, abdominal, and lower extremity regions, single or multiple and usually begin as a small area of edema that progress to local erythema and to hemorrhagic bullae, and finally to nerosis(Koriech and Al-Dash, 1988).

The characteristic histopathologic changes associated with generalized *Pseudomoas aeruginosa* infection are especially prominent in the skin lesions of ecthyma gangrenosum. There is usually evidence of hemorrhagic vascular necrosis with collection of gram-negative rods in the vessel walls. There is vasculitis with thrombosis, but often leukocytic infiltration around blood vessels is lacking(Dorff et al., 1971).

Prompt diagnosis and treatment are vital in the management of Pseudomonas infection. If the presence of ecthyma gangrenosum is suspected, appropriate cultures and biopsy specimens of the lesion should be obtained, as well as three blood cultures for general bacterial and sensitivity testing. Systemic antibiotic therapy should be started immediately, without waiting for laboratory confirmation. Several regimens are available, and the choice may depend on the usual pattern of antibiotic sensivity seen in the hospital. One such regimen consists of gentamycin, 1.5mg per kg of body weight given intravenously every eight hours, and carbenicillin 5 gm, or piperacillin 3 gm intravenously every four hours. If the organism is strongly suspected to be resistant to gentamycin, then a combination of carbenicillin 5 gm, or piperacillin 3 gm intravenously every four hours, and amikacin, 5mg per kg per dose, should be given intravenously every eight to 12 hours. Further adjustment of the antibiotics and the dosage can be done when the results of cultures and sensivity testing are known(Greene et al., 1984b). Early appropriate antiboitic therapy is necessary to prevent fatal invasive sepsis in ecthyma gangrenosum.

Our patient had underlying aplastic anemia with subsequent immunosuppressant therapy that left her with an impaired ability to respond to overwhelming infection. This is consistent with the findings of Valdivieso(1976). Thus the Pseudomonas septicemia that developed after the laparoscopic appendectomy probably led to the characteristic skin lesions of ecthyma gangrenosum and ultimate death despite the intensive antibiotic therapy. There has been a previous report in Korea(Lee et al., 1988), however this is the first case of ecthyma gangerenosum associated with aplastic anemia.

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