ECTHYMA GANGRENOsum: A RARE CUTANEOUS MANIFESTATION CAUSED BY PSEUDOMONAS AERUGINOSA WITHOUT BACTERAEMIA IN A LEUKAEMIC PATIENT- A CASE REPORT

*TN Singh, KM Devi, KS Devi

Abstract

Ecthyma gangrenosum is a rare and invasive cutaneous infection caused by Pseudomonas aeruginosa in the majority of cases, typically affecting immunocompromised patients, particularly those with neutropenia. We report a rare case of ecthyma gangrenosum in the absence of bacteraemia presenting as a solitary necrotic ulcer in a female patient with acute lymphoblastic leukaemia. A culture from the ecthyma lesion revealed the presence of Pseudomonas aeruginosa, but the results of repeated blood cultures were negative. The patient responded well to amikacin to which the isolate was susceptible in vitro. Considering high rate of mortality, early diagnosis and prompt effective treatment is mandatory.

Key words: Ecthyma gangrenosum , Neutropenia, Pseudomonas aeruginosa, RIMS

Ecthyma gangrenosum (EG) is a rare cutaneous lesion of systemic infection caused most commonly by Pseudomonas aeruginosa and observed in neutropenic patients. It may also rarely develop due to Pseudomonas aeruginosa in absence of bacteraemia.1 This disease had been related to life-threatening septicaemic infections and high mortality.2 It typically involves the extremities, gluteal and perineal regions. The predisposing factor that can lead to ecthyma gangrenosum is the presence of any kind of immunodeficiency associated with severe neutropenia.3 Most cases of ecthyma gangrenosum have been associated with concomitant septicaemia / bacteraemia. However, we report a rare case of non-septicaemic form of ecthyma gangrenosum presenting as a solitary necrotic ulcer in a patient with acute lymphoblastic leukaemia.

We isolated Pseudomonas aeruginosa from the ecthyma lesion in this patient. This is the first report of ecthyma gangrenosum caused by Pseudomonas aeruginosa from our hospital. This case is being reported herein to alert other clinical microbiologists and clinicians to avoid septicaemic life-threatening complications of ecthyma gangrenosum.

Case Report

A 21 years old female was admitted in RIMS hospital after experiencing five days of severe pain and two days of a necrotic lesion on the tip of left little toe. Pain and bluish colouration of the left little toe occurred following injury while cutting the nail. There was slight oozing of blood at the time of injury. On the next day, she became febrile and the following day she was noted to have a warm, raised, tender and indurated lesion on the tip of the left little toe. The lesion progress over the next two days to a haemorrhagic bluish bulla that ruptured to form a central area of necrosis surrounded by an erythematous halo. Blood cultures drawn at this time were repeatedly negative: Gram-stained smears from the necrotic lesion and from the surgical sample showed no pus cells but occasionally gram negative bacilli were seen. Cultivation of the pus swab revealed growth of Pseudomonas aeruginosa identified by conventional method.

On physical examination, the patient was well developed with a toxaemic appearance. Oral temperature was 39.5°C, respiratory rate 22 per minute, pulse rate 80 per minute, BP was 130/84 mm Hg. Chest X-ray was normal. Respiratory and cardiovascular systems were within normal limits. Abdomen was soft with normal bowel sound. The liver and spleen were not palpable. Central nervous system was normal. The patient was well oriented to time, place and person. Local examination of the ulcer: situated on the tip of left little toe, size: 1.5cm x 1.7cm with irregular shape, ragged undermined margins, raised hyperpigmented border and slough filled floor. Lesion was tender and the base was indurated.

The laboratory findings (at the time of admission) were: peripheral WBC count 85,000/cumm of which 1% were neutrophils 6% lymphocytes and 0% eosinophils and monocytes, haemoglobin was 10g/dL, platelet count 35,000/ cumm, ESR 110mm/hour, blast cells were 93% of which 48% were of hand mirror type, bleeding time was 1 minute 36 seconds and clotting time 4 minutes 30 seconds. The serum uric acid was 6.1 mg/dL, sodium 140 meq/L and potassium 4.3 meq/L. The blood concentration urea was 16mg/dL with serum creatinine 0.6 mg/dL, glucose 93 mg/dL (fasting) and 102 mg/dL (post prandial). Ultrasonography of the whole abdomen showed mild splenomegaly. The bone marrow

*Corresponding author (email: snabakr@rediffmail.com)
Department of Microbiology, Regional Institute of Medical Sciences(RIMS), Imphal-795 004, Manipur, India
Received: 28-09-04
Accepted: 10-03-05
analysis revealed the features of acute lymphoblastic leukaemia (L2 subtype). The antibiotic treatment was initiated with amikacin, to which the isolate was susceptible. This resulted in defervescence and clearance of *Pseudomonas aeruginosa*. The patient underwent surgical debridement of the necrotic lesion. The skin lesion resolved over a 21-day period. She was treated with intravenous doxorubicin and vincristine. Oral prednisolone and intrathecal methotrexate alternating with subcutaneous cytarabine were also administered. After the successful second dose of combination chemotherapy, the peripheral WBC count was 3250/cumm (28% neutrophils, 60% lymphocytes, 4% eosinophils and 6% monocytes), haemoglobin 10.4 g/dL, and platelet count was 60,000/cumm. She is currently under observation.

**Discussion**

Ecthyma gangrenosum is a characteristic dermatologic manifestation of severe and invasive infection caused most commonly by *Pseudomonas aeruginosa*. However, in some cases, it may be caused by *Klebsiella pneumoniae* and other species of *Pseudomonas* like *Pseudomonas maltophilia*, *Pseudomonas burkholderia*, *Pseudomonas cepacia* etc. Most cases of ecthyma gangrenosum have been associated with concomitant septicaemia but it may also rarely develop without bacteraemia due to *Pseudomonas aeruginosa*. Ecthyma gangrenosum is a well described entity which may occur with a frequency of 30% during the course of *Pseudomonas aeruginosa* septicaemia. The characteristic clinical appearance is red macules that progress to a central area of necrosis surrounded by an erythematous halo. This lesion represents a formidable skin sign of a potentially life-threatening systemic infection. The main site of ecthyma gangrenosum lesions is the gluteal or perineal region (57%), although this lesion can spread to other body sites, in which metastatic lesions appear on both trunks and lower extremities as was the case in our patient. Another major clinical feature in almost all patients is the presence of neutropenia. This disease is a life-threatening septicemic infection and has a high mortality. To decrease the mortality of this disease, the treatment should include prompt recognition of the skin lesion, appropriate antibiotic coverage against *Pseudomonas aeruginosa* and surgical debridement. The therapeutic intervention in our case was the empirical use of combination chemotherapy along with nutritional support.

The exact mechanism of the pathogenesis of ecthyma gangrenosum caused by *Pseudomonas aeruginosa* in neutropenic patients is poorly defined. The primary inciting factor appears to be the presence of numerous viable organisms at the point of involvement. Dissolution of the elastic lamina of blood vessels by *Pseudomonas* elastase allows for liberation of the bacilli into the subcutaneous tissues. Further prolific multiplication of the organism in subjacent tissue with elaboration of exotoxin A and proteases leads to the ulcerative lesion which is characterized by haemorrhage, encircled by a rim of reactive erythema. This cutaneous manifestation associated with *Pseudomonas aeruginosa* exotoxin A production has also been reported by Young *et al.*

In neutropenic patients the clinicians should be aware of such skin manifestations and consider ecthyma gangrenosum as a likely diagnosis to avoid the life-threatening septicemic infection and mortality.

**References**