



## Ecthyma gangrenosum in a previously healthy child

Adem Karbuz, Halil Özdemir, Gülsüm Kadioğlu Şimşek, Ergin Çiftçi, Erdal Ince, Ülker Doğru

Ankara University, Medical Faculty, Division of Pediatric Infectious Diseases, Ankara, Turkey

### Summary

Ecthyma gangrenosum is a well known cutaneous manifestation of *Pseudomonas aeruginosa* infections and is usually seen in immunocompromised patients. A previously healthy 5-month-old boy presented to our hospital with a complaint of an open inguinal wound. Physical examination revealed several ulcers surrounded by erythematous halos in bilateral inguinal regions. *Pseudomonas aeruginosa* was isolated from swab cultures obtained from the ulcers. However, blood cultures were sterile. The patient was diagnosed as non-bacterial ectyma gangrenosum. No immunological abnormality was detected during extensive investigation of the patient. The patient was successfully treated with an anti-pseudomonal antibiotic cefoperazone-sulbactam. (*Turk Arch Ped* 2013; 48: 68-70)

**Key words:** Ecthyma gangrenosum, *Pseudomonas aeruginosa*

### Introduction

Ecthyma gangrenosum is characterized by single or multiple ulcerated lesions in the skin or mucous membranes and occurs due to involvement of the skin by the pathogenic agent directly or due to hematogenous invasion (1,2). It is a well defined cutaneous finding of *Pseudomonas aeruginosa* infections especially in immunocompromised individuals. Ecthyma gangrenosum accompanies septicemia which may be fatal in most cases. However, rarely, it may occur due to *Pseudomonas aeruginosa* without bacteriemia and in previously healthy individuals (3,4,5,6).

### Case

A five-month-old male patient presented with open wounds in both inguinal regions. His medical history revealed that the lesions started one week before as erythematous, round, flat lesions two in the right inguinal region and one in the left inguinal region. The lesions were noted to be filled with blood in one day, the patient was presented to a health care institution and the lesions were drained there, but the patient was referred to us because the drained lesions deepened. The personal and familial history of the patient was normal. His physical growth was within normal limits. His

vital signs were found to be normal. Lesions ulcerated in the middle and surrounded by an erythematous halo two in the right inguinal region and one in the left inguinal region were observed (Picture 1 and 2). Laboratory findings were as follows: white blood cells 11 200/mm<sup>3</sup>, absolute neutrophil count 1 340/mm<sup>3</sup>, Hb 10,3 g/dL, platelets 400 000/mm<sup>3</sup>, erythrocyte sedimentation rate 18 mm/h, C-reactive protein 1.24 mg/dL (normal 0-0.8 mg/dL). 12% polymorphonuclear leukocytes, 68% lymphocytes, 16% monocytes and 4% atypical cells were observed on peripheral blood smear. Blood biochemical variables and complete urinalysis were found to be normal. The patient was internalized in the Pediatric Infectious Diseases Ward with a prediagnosis of ecthyma gangrenosum. After samples taken from the ulcerated lesions and blood were cultivated intravenous cefoperazone-sulbactam and clindamycin treatment was started. When it was learned that cultures taken from the lesions were positive for *P. aeruginosa*, clindamycin treatment was discontinued. Hemocultures were sterile. The patient was diagnosed as non-bacteriemic ecthyma gangrenosum. The immunological investigations of the patient who had no known underlying disease were found within normal limits: IgA: 0.474 g/L (0.18-0.5), IgM: 0.872 g/L (0.45-2), IgG: 6.06 g/L (4.2-12) and lymphocyte subgroups were found to be normal. Blastic change was found to be normal and nitro-blue

tetrazolium test which was done for chronic granulomatous disease was found to be 100%. The patient whose lesions healed completely in the follow-up was discharged after a treatment period of 10 days.

## Discussion

Ecthyma gangrenosum is a very rare vasculitis in which the adventitia and media of the venous cutaneous blood vessels are affected and the intima and lumina are kept intact (7). This may occur via the blood or inoculation of the skin directly (2,3). However, the arterial blood vessels are not affected (7).

Ecthyma gangrenosum starts as painless, red macules and advances to papules and hemorrhagic bullae. When these lesions are opened, gangrenous ulcers with grey-black scars surrounded by an erythematous halo appear (1,4,5). The lesions may mature in a short period (in 12 hours) or may transform into ulcers with necrosis in the middle in a few days and they may be in different



Picture 1. Two lesions ulcerated in the middle and surrounded by an erythematous halo in the right inguinal region



Picture 2. A lesion ulcerated in the middle and surrounded by an erythematous halo in the left inguinal region

development stages (2,6). The history of our patient revealed that the lesions followed the typical development stages. The lesions are generally multiple both in septic and non-septicemic cases (4). The localizations of the lesions are as follows: 57% gluteal and perianal regions, 30% extremities, 6% trunk and 6% face (2,3).

It is accepted that ecthyma gangrenosum is mostly related with infections caused by *P. aeruginosa* (2). In addition, it has been found that clinically similar lesions can develop with *Staphylococcus aureus*, *Aeromonas hydrophilia*, *Enterobacter species*, *Proteus species*, *Burkholderia cepacia*, *Serratia marcescens*, *Aspergillus species*, *Mucor species*, *Escherichia coli* and *Candida species* (7). Kanehiro et al. (8) reported that 60% of ecthyma gangrenosum cases in Japan were related with staphylococcus and the remaining were related with streptococcus and *Pseudomonas* infections with decreasing frequency. There are histological differences between ecthyma gangrenosum caused by *Pseudomonas aeruginosa* and ecthyma gangrenosum caused by streptococci and staphylococci. Skin biopsy was considered for the lesions of our patient, but it was canceled, when the culture was found to be positive for *P. aeruginosa* and the lesions healed with the treatment administered.

*Pseudomonas aeruginosa* is an aerobic gram negative bacterium and leads to opportunistic infections especially in patients who have underlying chronic disease and whose immune systems are affected (5). Ecthyma gangrenosum may occur during systemic *P. aeruginosa* infections or as primary skin lesion via direct inoculation (7). Ecthyma gangrenosum can be examined as two types according to its pathogenesis. Classical ecthyma gangrenosum is bacteriemic; the pathogenic microorganism in the circulation (frequently *P. aeruginosa*) reaches the skin via the hematogenous route (4). In classical ecthyma gangrenosum, the lesions usually occur in the plicae (7). The mortality rate related with *Pseudomonas* septicemia in immunocompromised patients ranges between 38% and 77% (2,9). The second type is more benign and can be named as regional, non-septicemic ecthyma gangrenosum; here, the pathogenic agent is inoculated directly into the skin. This type is observed in infants in whom immune deficiency could not be diagnosed before or who have transient risk factors for development of ecthyma gangrenosum. In this type, a few necrotic ulcers occur mostly in the buttocks (6). The mortality rate is 15% in this type of ecthyma gangrenosum which is non-bacteriemic and generally involves negative blood culture (2,10).

*Pseudomonas aeruginosa* infections are closely related with the defense mechanisms of the host and the most important defense factor is neutrophils. Quantitative and qualitative disorders of neutrophils constitute a significant risk for development of ecthyma gangrenosum. Most of the previously healthy individuals who develop ecthyma

gangrenosum are neutropenic (2). Although neutropenia is a significant risk factor, *Pseudomonas* infections may lead to transient neutropenia during the disease by inhibiting neutrophil migration in the involved area and by decreasing the neutrophil count in the circulation via toxins (2,11). In addition, patients who have viral infection, who have a history of use of wide-spectrum antibiotics and most importantly who have chronic disease and immune deficiency are under risk in terms of *Pseudomonas* infections (2). Although there are case reports which show that ecthyma gangrenosum occurs in previously healthy children, a review stated that most patients had a risk factor for development of ecthyma gangrenosum or an underlying medical condition which had not been elucidated yet (6). Therefore, in many resources, it is emphasized that a complete immunological evaluation should be performed in previously healthy individuals with ecthyma gangrenosum (2,5,6). The necessity of performing complete blood count in terms of cyclic neutropenia, transient neutropenia or chronic neutropenia after the disease has also been emphasized (2). However, community-acquired *P. aeruginosa* infection may occur in healthy infants below the age of one (5,11,12). Our patient had no history of recent viral infection or use of antibiotics or an underlying disease. He only had a mild neutropenia. A complete immunological examination was performed in our patient in terms of an underlying deficiency, but no problem was found. Follow-up examinations performed afterwards were also completely normal. The mild neutropenia in our patient was probably a transient neutropenia developed during *Pseudomonas* infection in relation with toxins.

In treatment, early diagnosis and use of appropriate antibiotics are important. An anti-pseudomonal beta-lactam antibiotic in combination with an aminoglycoside can be started blindly for bacteriemic and non-bacteriemic ecthyma gangrenosum (4). Afterwards, antibiotic treatment may be changed according to the antibiotic sensitivity results (1). In our patient, sefoperazone-sulbactam for *Pseudomonas* infection and clindamycin for group A streptococcal and staphylococcal infections were started initially. When it was learned that the culture taken from the lesion was positive for *P. aeruginosa*, clindamycin treatment was discontinued and

the treatment was completed to 10 days with sefoperazone-sulbactam to which the pathogenic agent was sensitive. The skin lesions closed in the follow-up and no problem was found in follow-up visits.

Conclusively, although ecthyma gangrenosum occurs very rarely in individuals who have no risk factor, non-bacteriemic ecthyma gangrenosum may be observed rarely. These patients should be investigated in terms of an underlying immune deficiency, even if they have been completely healthy previously.

## References

1. Athappan G, Unnikrishnan A, Chandraprakasam S. Ecthyma gangrenosum: presentation in a normal neonate. *Dermatol Online J* 2008; 14(2):7.
2. Zomorodi A, Wald ER. Ecthyma gangrenosum: considerations in a previously healthy child. *Pediatr Infect Dis J* 2002; 21(12): 1161-1164.
3. Duman M, Özdemir D, Yiş U, Köroğlu TF, Ören O, Berktaş S. Multiple erythematous nodules and ecthyma gangrenosum as a manifestation of *Pseudomonas aeruginosa* sepsis in a previously healthy infant. *Pediatr Dermatol* 2006; 23(3): 243-246.
4. Song WK, Kim YC, Park HJ, Cinn YW. Ecthyma gangrenosum without bacteraemia in a leukaemic patient. *Clin Exp Dermatol* 2001; 26(5): 395-397.
5. Chan YH, Chong CY, Puthuchery J, Loh TF. Ecthyma gangrenosum: a manifestation of *Pseudomonas* sepsis in three paediatric patients. *Singapore Med J* 2006; 47(12): 1080-1083.
6. Ishikawa T, Sakurai Y, Tanaka M, Daikoku N, Ishihara T, Nakajima M, Miyagawa S, Yoshioka A. Ecthyma gangrenosum-like lesions in a healthy child after infection treated with antibiotics. *Pediatr Dermatol* 2005; 22(5): 453-456.
7. Morelli JG. Cutaneous bacterial infections. In: Kliegman RM, Behrman RE, Jansen HB, Stanton BF, (eds). *Nelson textbook of pediatrics*. 18th ed. Philadelphia: Saunders, 2007: 2740.
8. Kanehiro A, Nishino M, Yoshioka A, et al. Two case reports of neonatal ecthyma gangrenosum with septicemia. *Jpn J Pediatr* 1986; 39: 179-184.
9. Altemeier WA, Todd JC, Inge WW. Gram-negative septicemia: a growing threat. *Ann Surg* 1967; 166(4): 530-542.
10. Huminer D, Siegman-Igra Y, Morduchowicz G, Pittlik SD. Ecthyma gangrenosum without bacteremia. Report of six cases and review of the literature. *Arch Intern Med* 1987; 147(2): 299-301.
11. Chusid MJ, Hillmann SM. Community-acquired *Pseudomonas* sepsis in previously healthy infants. *Pediatr Infect Dis J* 1987; 6(7): 681-684.
12. Mull CC, Scarfone RJ, Conway D. Ecthyma gangrenosum as a manifestation of *Pseudomonas* sepsis in a previously healthy child. *Ann Emerg Med* 2000; 36(4): 383-387.