A Case of Neutrophilic Eccrine Hidradenitis in an Infant

Kyoung Jin Kim, M.D., Jeong Yeob Lee, M.D., Jee Ho Choi, M.D.,
Kyung Jeh Sung, M.D., Kee Chan Moon, M.D., Jai Kyoung Koh, M.D.

Department of Dermatology, Asan Medical Center, College of Medicine, University of Ulsan, Seoul, Korea

Neutrophilic eccrine hidradenitis (NEH) is a rare neutrophilic dermatosis which is characterized by the erythematous papules or plaques with a neutrophilic infiltrates around eccrine glands and coils. In 90% of cases, NEH develops in patients who have received chemotherapy for malignant diseases, but other drugs, infections, and paraneoplastic phenomenons are considered as possible etiologic factors. Furthermore, NEH is known to be extremely rare in infants. Herein we report a case of NEH in a nine-month-old infant with acute myelogenous leukemia (AML), which lesions appeared on the extremities including palms and soles before starting chemotherapy. (Ann Dermatol 14(2) 106-109, 2002).

Key Words : Neutrophilic eccrine hidradenitis (NEH), Infant, Before chemotherapy

Neutrophilic eccrine hidradenitis (NEH) is a relatively rare neutrophilic dermatosis and first described as a transient and benign complication of various chemotherapy regimens for acute leukemia. More recently, NEH has been observed in other conditions, such as infections or drugs, before the diagnosis of malignancy and even generally healthy individuals. Furthermore, there were several reports of idiopathic palmoplantar hidradenitis, which had similar clinical and histopathologic findings with NEH, in children without history of chemotherapy.

We herein describe a nine-month-old female infant with NEH on the extremities including palms and soles. Although she had acute myelogenous leukemia (AML), the lesions of NEH appeared before starting chemotherapy, suggestive of other etiology.

Received June 18, 2001.
Accepted for publication November 7, 2001.
Reprint request to : Jee Ho Choi, M.D., Department of Dermatology, Asan Medical Center, College of Medicine, University of Ulsan
388-1 Poongnap-dong, Songpa-gu
Seoul, 138-736 Korea
Tel. (02)3010-3460, Fax. (02)486-7831
E-mail: kimkj20@dreamwiz.com

CASE REPORT

A nine-month-old female infant presented with multiple, erythematous, tender maculopapules and patches on the upper and lower extremities for two months. Two months earlier, cough and easy bruisability developed and several erythematous infiltrative papules appeared on both dorsum of hands and feet one week later. Thereafter fever, decreased appetite, and dyspnea presented. She was diagnosed as a pneumonia with sepsis and treated with antibiotics, but there was no clinical improvement. The skin lesions were more erythematous and wide spread to both forearms, legs, palms and soles. Laboratory investigations showed the following results or findings: leukocytes, $31.2\times 10^3/mm^3$ (normal, 4.0 to $10.0\times 10^3/mm^3$), with differential count of monocytes, 20%, lymphocytes, 21%, atypical lymphocytes, 1%, segmental neutrophils, 49%, hemoglobin, 10.9 g/dl (normal, 12 to 16 g/dl), platelet, $170\times 10^3/mm^3$ (normal, 150 to $350\times 10^3/mm^3$), CRP, 19.11mg/dl (normal, 0-0.6), reticulocytes, 46.5% (normal, 0 to 17%), and peripheral blood smear showed immature and large dysplastic monocytes, suggestive of myelodysplastic syndrome. A bone marrow biopsy was done and she was diagnosed as AML, M4.

The erythematous skin lesions increased in number

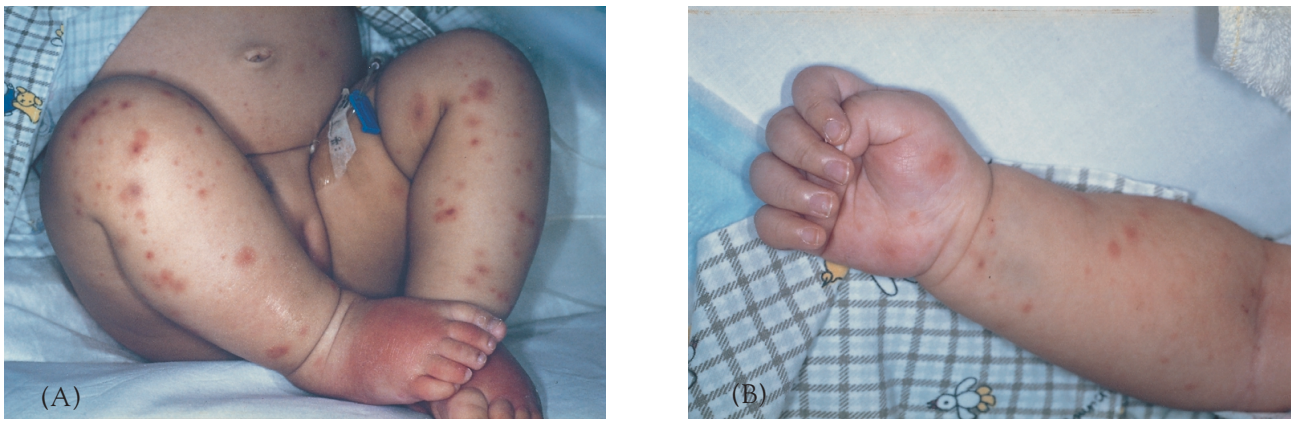

Fig. 1. Multiple, tender, erythematous to purpuric macules, papules and patches on (A) both lower extremities and (B) forearm including palm.

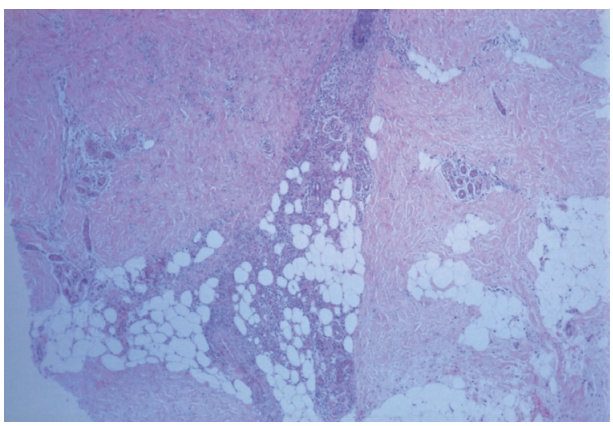

Fig. 2. A dense linearly distributed nodular neutrophilic infiltrate around eccrine sweat units extends to the subcutis (H&E, ×40).

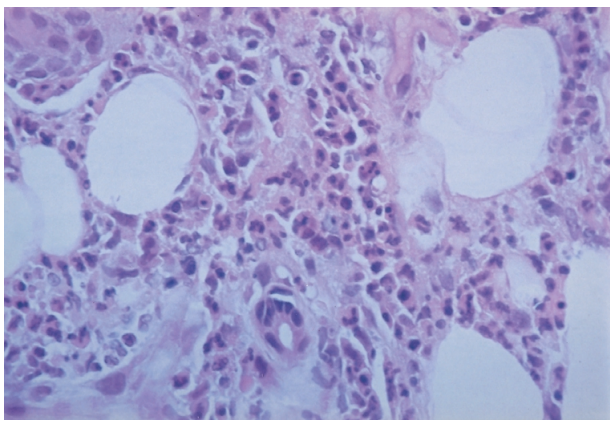

Fig. 3. Neutrophilis around the eccrine glands (H&E, ×400).

and the patient was referred to the dermatologic clinic. Skin examination revealed multiple, slightly tender, erythematous or some purpuric, discrete macules, papules, and patches on the both upper and lower extremities including palms and soles (Fig. 1). A skin biopsy showed an unremarkable epidermis but a dense linear and nodular infiltrate composed predominantly of neutrophils that was localized to the eccrine sweat units with slight extension to the neighboring periglandular area and subcutis (Fig. 2 and 3). Also, there was focal necrosis of eccrine coils. Apart from a mild mixed perivascular infiltrate in the middle and deep dermis, no other significant changes were observed. These clinical and histopathologic features enabled diagnosis of neutrophilic eccrine hidradenitis to be made.

One day after biopsy, chemotherapy regimen composed of cytosine arabinoside and doxorubicin was started. She tolerated well to the chemotherapy and cutaneous lesions had completely disappeared after two weeks without recurrence for follow-up period of six months.

DISCUSSION

Since first being described by Harris et al.[1] in a patient with chemotherapy using cytarabine and doxorubicin for AML in 1982, approximately 50-60 cases of NEH have been reported in literatures[2]. There is slight male predominance and mean age is 40.3 years (1-79 years). Besides only one case of NEH developed in a one-year-old baby, there has

been no description about infantile NEH to our knowledge. Our patient was a nine-month-old infant and we think that she may be the youngest patient with NEH in literature.

Most common is the leukemic patient receiving systemic chemotherapy, but NEH has also been documented in individuals undergoing chemotherapy for a number of other types of cancer[2,3]. Furthermore, there was some reports of NEH without association of chemotherapy. These include the infectious NEH (HIV, serratia marescens, Enterobacter cloacae, Staphylococcus aureus), drug-induced NEH (acetaminophen, G-CSF, zidovudine), and paraneoplastic NEH[2,4-8]. Pierson et al.[9] described a patient with NEH arising several weeks prior to the onset of fulminant AML and insisted that the NEH might develop as a paraneoplastic phenomenon in their case. Although our patient had suffered from AML, because the lesions developed before chemotherapy, we might speculate that upper respiratory infection or medication might be a possible cause or NEH could be a paraneoplastic phenomenon in our case.

The cutaneous lesions of NEH are featured by the development of infiltrative or edematous papules or plaques, asymptomatic or painful, close to Sweet's syndrome[2]. The pigmented or purpuric lesions have also been reported. They can develop either in a proximal disposition involving the upper trunk, upper limbs and the face, particularly the periorbital areas, but distal extremities including palms and soles are rarely involved[2]. Moreover, when the lesions limited to the palms and/or soles with clinical and histopathologic resemblance with NEH in a child without history of chemotherapy or malignancy, the term of "idiopathic palmoplantar hidradenitis" is used[3,10]. Since our patient had involvement of the palms and soles similar to idiopathic palmoplantar hidradenitis but also leukemic symptoms, we believe that these features are those of true NEH.

The definite diagnosis of NEH relies on the histopathologic examination[2]. The hallmark of this condition is characterized by the association of a degeneration of eccrine glands, and dense neutrophilic infiltrate surrounding the eccrine coils and into the eccrine epithelium. Other findings including diffuse dermal edema, hemorrhage, diffuse or perivascular lymphohistiocytic infiltration can also be observed.

The pathogenesis of NEH is not yet known. Two major hypotheses, however, are proposed in the current literatures[2]. One is a possible direct toxic effect on the eccrine apparatus and this may ultimately be shown to account for many of the chemotherapy-related cases. The other is that NEH is a hypersensitivity reaction within the spectrum of neutrophilic dermatoses such as Sweet's syndrome. The minor explanation for pathogenesis of NEH include the suggestion that neutrophilic infiltrate can be secondary to sweat gland abnormalities or a paraneoplastic phenomenon[2,8].

No specific treatment is required and spontaneous resolution of lesions of NEH without scarring is always observed within a few days or weeks, but relapse can occur in some patients receiving the same chemotherapy regimen. Antibiotics and nonsteroidal anti-inflammatory drugs are used for shortening of duration and relief of pain. Dapsone can be used for prevention of recurrent NEH in a patient receiving repeated chemotherapy[11].

Several aspects from our patient are worth noting. First, the patient was nine-month old, the youngest age among which has been reported. Secondly, the lesions involved the relatively uncommon sites of distal extremities including palms and soles. Finally, NEH developed before chemotherapy, suggestive of other etiologic factors such as infection, drug, or malignancy itself.

REFERENCES

1. Harrist TJ, Fine JD, Berman RS, et al: Neutrophilic eccrine hidradenitis. A distinctive type of neutrophilic dermatosis associated with myelogenous leukemia and chemotherapy. Arch Dermatol 118:263-266, 1982
2. Bachmeyer C, Aractingi S: Neutrophilic eccrine hidradenitis. Clinics in Dermatology 18:319-330, 2000
3. Stahr BJ, Cooper PH, Caputo RV: Idiopathic plantar hidradenitis: a neutrophilic eccrine hidradenitis occurring primarily in children. J Cutan Pathol 21:289-296, 1994
4. Moreno A, Barnadas MA, Ravella A, DeMoragas JM: Infectious eccrine hidradenitis in a patient undergoing hemodialysis. Arch Dermatol 121:1106, 1985
5. Allegue F, Rocamora A, Martin-Gonzalez M, Alonso ML, Ledo A: Infectious eccrine hidradenitis. J Am Acad Dermatol 22:1119, 1990

6. Bachmeyer C, Chaibi P, Aractingi S: Neutrophilic eccrine hidradenitis induced by granulocyte colony-stimulating factor. Br J Dermatol 139:340-341, 1998
7. Bachmeyer C, Reygagne P, Aractingi S: Recurrent neutrophilic eccrine hidradenitis in an HIV-1-infected patient. Dermatology 200:328-330, 2000
8. Kuttner BJ, Kurban RS: Neutrophilic eccrine hidradenitis in the absence of an underlying malignancy. Cutis 41:403-405, 1988
9. Pierson JC, Helm TN, Taylor JS: Neutrophilic eccrine hidradenitis heralding the onset of acute myelogenous leukemia. Arch Dermatol 129:791-792, 1993
10. Rabinowitz LG, Cintra ML, Hood AF, Esterly NB: Recurrent palmoplantar hidradenitis in children. Arch Dermatol 131:817-820, 1995
11. Shear NH, Knowles SR, Shapiro L, et al: Dapsone in prevention of recurrent neutrophilic eccrine hidradenitis. J Am Acad Dermatol 35:819-822, 1996