A Case of Early Age Onset Hailey-Hailey Disease Treated with Surgical Operation

Yong Sub Oh, M.D., Sung Yul Lee, M.D., Hae Joon Song, M.D.,
Young Chul Kye, M.D., Soo Nam Kim, M.D.

Department of Dermatology, College of Medicine, Korea University Seoul, Korea

Hailey-Hailey disease is a rare hereditary dermatosis that begins in the 2nd or 3rd decade of life. The skin lesion is characterized by a localized, recurrent eruption of small vesicles on an erythematous base. It courses remissions and exacerbations. It seldom begins in early childhood, and main treatment modalities are conservative ones.

We report a case of Hailey-Hailey disease that began on a 7-month old infant and improved by surgical treatment. In according to review of the previous reports, it is probably the earliest onset age and it is may be the first case which was treated with surgery in Korea. (Ann Dermatol 6:(1) 86~89, 1994)

Key Words: Hailey-Hailey disease, 7-month old age, Split thickness skin graft.

Hailey-Hailey disease is a rare hereditary dermatosis that was first described in 1939[1]. Usually, it begins in the 2nd or 3rd decade of life. In only rare instance does it start in early childhood.[2] Characteristic skin lesions are localized, recurrent eruption of very small vesicles on the erythematous base and exudative lesions involving the neck, groin, axilla and intertriginous areas. The exacerbations may be precipitated by a warm, humid environment, friction, mechanical trauma, UV radiation and bacterial or fungal infections.[3]

We report a case of Hailey-Hailey disease that began on a 7-month male infant and was successfully treated with excision and split thickness skin graft.

REPORT OF A CASE

A 7 month-old Korean child was presented with a left popliteal skin lesion that had developed 4 months earlier. The lesion was a erythematous plaque with small peripheral spreading vesicles. It enlarged with erosion, fissure, and ulceration progressively (Fig.1). His familial and past medical history was non-contributory.

On physical examinations the patient was relatively healthy. Routine laboratory studies including complete blood count, urine analysis, liver function test, VDRL were within normal limits. X-ray film showed no abnormalities in the chest and EKG was normal. Direct wound cultures showed positive for Group D streptococcus, but negative for fungus.

The skin biopsy specimen was obtained from the left popliteal skin lesion for histology. The epidermis showed a suprabasal cleft formation with extensive acantholysis. The villi were protruded into the bulla cavity but dilapidated brick wall appearance and dyskeratotic figures were not shown. The dermis showed inflammatory cell infiltrates (Fig. 2).

Direct and indirect immunofluoresence studies were negative.

Electron microscopic findings showed that bizarre microvilli changed on the surface of keratinocytes and the tonofilaments became thickened and arranged in a perinuclear fashion (Fig. 3).

We started dapsone and erythromycin with top-

Received March 13, 1993
Accepted for publication August 12, 1993
Reprint request to: Young Sub Oh M.D., Department of Dermatology, College of Medicine, Korea University Seoul, Korea

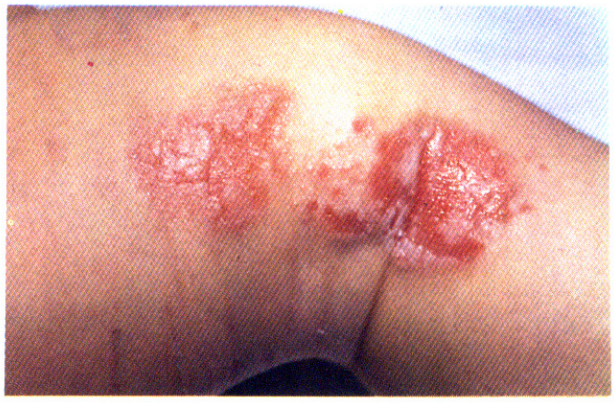

Fig. 1. Erosion, fissure, ulceration with small peripheral spreading vesicles on the erythematous base on popliteal area.

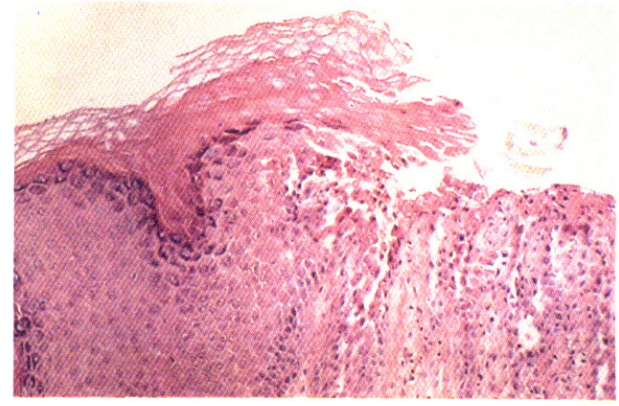

Fig. 2. Suprabasal acantholysis with blister formation was showed and the villi were protruded into the bulla cavity (H & E stain, ×100)

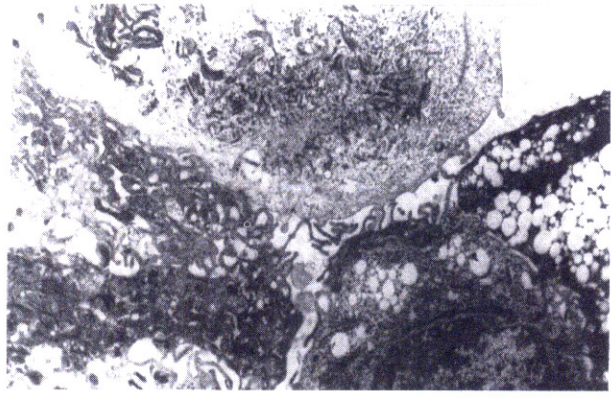

Fig. 3. Electron micrograph. The bizarre microvilli changed on the surface of keratinocytes and thickened tonofilaments around the perinuclear region. (×5,000)

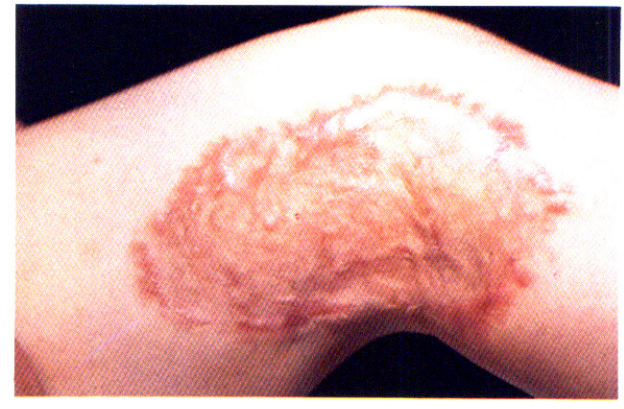

Fig. 4. After split thickness skin graft, the graft site was healed well. Although there was some hypertrophic scar peripherally, the recurrence was not revealed up to date.

ical corticosteroid for 5 weeks, but improvement was not noticed. So we treated the patient with excision and split thickness skin graft.

Although there was a hypertrophic scar peripherally after surgery, the graft site healed well. No evidence of recurrence was revealed up to date (Fig. 4).

DISCUSSION

Hailey-Hailey disease occurs usually just after puberty, affecting primarily young adults with the highest prevalence in the 15 to 30 age group, and both sexes are equally affected. Kandhari et al[2] reported Hailey-Hailey disease developed in an Indian boy aged 4 and 1/2 years. A family history of the disease is present in about two thirds of the cases, and the condition is considered to be inherited as an autosomal dominant gene with irregular penetrance.[4] The general health is not usually affected. However itching and burning are common symptoms. In addition, maceration and fissuring in the intertriginous areas commonly causes pain on motion. The clinical course is characterized by spontaneous exacerbations and remissions but there is little or no tendency to improvement with age.[5]

Histopathologic findings of Hailey-Hailey disease show a gradual progression from suprabasilar lacuna in early lesions to large suprabasilar acantholytic vesicles and bullae. The villi consist of elongated papillae covered by one or several layers of keratinocytes and it protrudes into the blister cavity. Intercellular edema with the loss of intercellular attachments leads to partial acantholysis that is referred to as a dilapidated brick wall ap-

pearance in the lower epidermis.[3,6]

The electron microscopic studies showed the detachment of tonofilaments from desmosomes, which resulted in retraction and clumping of the tonofilaments, degeneration of the desmosomes and subsequent acantholysis[3,7,8]. Gottlieb and Lutzner[7] emphasized that the combination of microvillose change of the keratinocytes and the abnormal tonofilament configuration that occurred in the same lesion was pathognomonic of Hailey-Hailey disease.

Generally, the results of direct and indirect Immunofluorescence studies are negative in Hailey-Hailey disease[3,5]

Hailey-Hailey disease may resemble impetigo, pemphigus vulgaris, pemphigus vegetans, Darier's disease and transient acantholytic dermatosis, but Hailey-Hailey disease is differentiated from other diseases by clinical and histopathologic findings.[3,5,6] The long duration and recurrent tendency, the location of lesion and histologic findings can lead to impetigo being rule out.[5] Generally immunofluorescence studies that are positive in pemphigus vulgaris and negative in Hailey-Hailey disease, allow for a definite distinction.[3,5] Differentiation from Darier's disease is not difficult in most instances, because in Darier's disease, histopathologic findings show smaller suprabasal separation, less pronounced acantholysis and much more dyskeratosis consisting of the formation of corps ronds and grains. In addition, Darier's disease usually distributes on seborrheic areas and has keratotic papules with follicular arrangement and is more persistent.[5,6] Transient acantholytic dermatosis resembles Hailey-Hailey disease histopathologically but a clinicopathologic correlation can lead to an accurate diagnosis.[3,5,9]

There are several treatment modalities for Hailey-Hailey disease.

At times, the use of topical and oral antibiotics and antifungal therapy result in notable improvement.[3,4,5] In some instances, dapsone has been found to be effective.[3,5] The systemic administration of corticosteroid is effective only in severe cases.[5] Topical application of corticosteroid creams or lotions is of limited value but may be used. The surgical operation, wide excision and split thickness skin graft, is used when medical and other therapeutic modalities have failed and the disease is disabling.[3,10] Although flare-up at the periphery of the graft can occur, the result of surgical operation has generally been gratifying. Surgical operation has attributed to the scar formation at the base of the graft, which leads to the destruction of the sweat glands and of the nerves innervating these glands. The destruction of these gives a results in alteration of the local environment which becomes more dry and gives a decreased opportunity for fungal and bacterial growth.[3] Other treatment modalities include, vit-E 800 to 1200 IU daily administration,[11] cryosurgery,[12] and electrodessication[13] have been previously reported but generally it is not accepted. Recently, it was reported that carbon dioxide laser vaporization was effective.[14]

In Korea, eleven cases have been reported since 1964.[9,15-21] Age range was 24 to 50 years, and the patients consisted of 9 male and 2 female patients. The predilection sites were neck, groin, and axillae. Only 3 cases had a clear family history. All were treated with medical therapies consisted of tetracycline and other susceptible antibiotics, corticosteroids, antihistamines, dapsone, topical antifungals, and wet dressing with KMn04 or Burrow solutions.

Our case begins on 7-month male infant with the skin lesion developed on his left popliteal area. There was no family history of Hailey-Hailey disease. We started dapsone and erythromycin with topical corticosteroid for 5 weeks but this failed. So finally we treated the patient with excision and split thickness skin graft and obtained a favorable outcome.

REFERENCES

1. Hailey H, Hailey H: Familial benign chronic pemphigus. Arch Dermatol 39 : 679-685, 1939.
2. Kandhari KC: Chronic benign pemphigus of Hailey and Hailey in an Indian child. Br J Dermatol 75 : 212, 1963.
3. Michael B: "Familial benign chronic pemphigus" by Hailey-Hailey, April 1939. Commentary : Hailey-Hailey disease. Arch Dermatol 118 : 774-783, 1982.
4. Moschella SL, Hurley HJ: Dermatology, 3rd ed, W. B. Saunders Co, Philadelphia, 1992, pp 687-689.
5. Fitzpatrick TB: Dermatology in general medicine. 3rd ed, McGraw-Hill book company, New York, 1987, pp 598-601.

6. Lever WF, Schaumburg-Lever G: Histopathology of the skin. 7th ed, J B Lipincott Co, Philadelphia, 1990, pp 82-83.
7. Gottlieb SK, Lutzner MA: Hailey-Hailey disease; An electron microscopic study. J Invest Dermatol 54 : 368-376, 1970.
8. Tak MJH, Park YK, Lee SN: Electron microscopic study of familial benign chronic pemphigus. Kor J Dermatol 20 : 777-779, 1982.
9. Kim KH, Kim EC, Lee AY, Cho KH, Lee YS: A case of Grover's disease showing chronic course. Kor J Dermatol 27: 582-587, 1987.
10. Menz T, Jackson IT, Connolly S: Surgical control of Hailey-Hailey disease. Br J Plastic Surg 40 : 557-561, 1987.
11. Ayres S jr: Hailey-Hailey diseas; Response to vitamin E therapy. Arch Dermatol 119 : 450, 1983.
12. Galimberti RL, Kowalczuk AM, Bianchi 0: Chronic benign familial pemphigus. Int J Dermatol 27 : 495-500, 1988.
13. Quitadamo MJ, Spencer SK: Surgical management of Hailey-Hailey disease. J Am Acad Dermatol 25 : 342, 1991.
14. Kartamaa M, Reitamo S: Familial benign chronic pemphigus, treatment with carbon dioxide laser vaporization. Arch Dermatol 128 : 646-648, 1992.
15. Suh CK: A case of familial benign chronic pemphigus. Kor J Dermatol 3 : 47-50, 1964.
16. Lee JB, Houh W: A case of familial benign chronic pemphigus. Kor J Dermatol 12 : 245-248, 1974.
17. Kim YP, Cho JH: Genetic observation of familial benign chronic pemphigus and report of two cases with review of literatures. Kor J Dermatol 13 : 61-66, 1975.
18. Choi YJ, Chung TA: Three cases of familial benign chronic pemphigus. Kor J Dermatol 17 : 379-381, 1979.
19. Byun JH, Kook HI: A case of familial benign chronic pemphigus. Kor J Dermatol 18 : 433-434, 1980.
20. Lee JS, Chyung EJ, Park SY: A case of familial benign chronic pemphigus. Kor J Dermatol 22 : 639-641, 1984.
21. Kim NI, Lee CB, Lee MH, Houh CL: A case of familial benign chronic pemphigus. Kor J Dermatol 25 : 716-718, 1987.