梨花醫大誌：第3卷·第1號·1980
Ewha Med. J. Vol. 3, No. 1, 1980

Two Cases of Subcorneal Pustular Dermatosis

Joo Hyun Byun, M.D., Jeong Hee Hahm, M.D., and Hong Il Kook, M.D.

Department of Dermatology, College of Medicine, Ewha Womans University.

=國文抄錄=

Subcorneal Pustular Dermatosis의 2例

梨花女子大學校 醫科大學 皮膚科學敎室

邊 宙 鉉·咸 妊 希·鞠 泓 一

角質下 膿疱性 皮膚病은 Sneddon과 Wilkinson에 依해서 처음으로 報告된 以來 많은 症例가 報告되어왔다. 이는 慢性的으로 小水疱, 膿疱의 發生이 대부분 反復的으로 好轉과 惡化를 이루며, 中年層의 女子에게서 흔히 볼 수 있는 疾患이다.

本敎室에서는 體幹部, 上·下肢에 水疱性發疹이 나타나면서 好轉과 惡化를 反復하였고, 內院 당시 대칭적으로 全身에 瘙痒症을 同伴한 環狀形 紅斑 주위에 針頭大의 膿疱疹 및 痂皮, 鱗屑, 剝皮가 發生하였으며, 組織所見으로 角質下 膿疱와 好中球浸潤을 보이는 2例를 經驗하였기에 報告하는 바이다.

Subcorneal pustular dermatosis, first described by Sneedon and Wilkinson in 1956[1], is a chronic, benigh, relapsing vesicopustular eruption, which involves the intertrigenous and flexual region, with an unknown etiology and pathogenesis.

This relatively rare disease occur most freqtently in middle aged women, but young adults and elderly persons of either sex can be affected. A number of cases in children have occurred[2)3)].

The treatment of choice is diaminodiphylsulphone (dapsone) or sulphapyridine: corticosteroids are less effective[4)]. Successful results of oral treatment with vitamin E[2)5)6)] and retinoic acid derivatives(Ro-9259) have been reported. Moreover, alternately retinoic acid and corticosteroid locally with success reported[7)].

In this paper we report two patients with subcorneal pustular dermatosis, including treatment with dapsone, corticosteroids and ultraviolet light producing satisfactory clinical response transiently, but a complete remission was not obtained.

Report of Cases

CASE 1. A 38-year-old woman was admitted to our department with pruritic recurrent pin head sized pustules with erythematous base on the trunk, both arms and thighs since 10 years ago. 10 years prior to visit our department, she first found miliary sized vesicles and pustules with erythematous base developing on the entire body in ninth month of gestation. She had admitted two times at certain hospital for the similar skin lesions respectively nine months and two months of gestation. At first admission, she was treated

with dapsone and corticosteroids for two months under the impression of herpes gestationis. After delivery, skin lesions did not recur. Two years later, she had suffered from abdominal pain and similar skin lesions reappeared at the eight weeks of gestation. On second admission, she was treated with corticosteroid for ten months. One month after discharge she could not walk for the pain of hip joint. Bone X-ray examination showed pathological fracture of femur head due to osteoporosis, which was resulted from prolonged therapy of corticosteroid. Similar skin lesions have used to occur at spring time every year.

On this admission, the physical examination revealed the patient has ill looking appearance. Circular and annular configurated maculopustular patches actively spread peripherally with sloughing on the upper arm and axillae. Confluented and discreted annular pustules with erythematous base on the abdomen, and stocking type of distribution of healed skin and advancing border of the active lesion of the upper thighs noted(Fig. 1, 2, 3).

The remainder of the physical examination was unremarkable.

Laboratory studies showed a hemoglobin level of 12.2 gm/dl and a hematocrit reading of 36% with normal indices. White blood cell count was 9,100/cu mm, with 70% neutrophils, 11% band forms, 18% lymphocytes, and 1% eosinophils.

Platelets was 210.000/cu mm; ESR, 54mm/hr, urinalysis, normal. Normal results were obtained for liver function test, and electrolytes. Chest X-ray was normal. Culture from the skin lesions grew staphylococcus aureus. KOH mount was negative. Punch biopsy specimen were taken from the left shoulder and upper arm. In H-E stain there were subcorneal pustules containing polymorphonuclear leukocytes and chronic inflammatory cell infiltration in the dermis (Fig. 4)

She was treated with dapsone, 50 mg, three times a day, and corticosteroid ointment following after 1:7,000 potassium permanganate bathing with mild effectiveness.

CASE 2. A 49-year-old female visited to our department with pruritic, recurrent, pin head to match head sized pustules with erythematous base developing on the both legs for eight years. Prior to visit our department, she first noticed the pustules were developed on the soles and extended to the dorsal area of feet. Four years to prior to visit, the pustules were flaccid and actively spread peripherally, forming circinate pattern on the both legs. Shs had experienced no significant illness in her past history.

The physical examination was unremarkable except skin lesion. Sloughing with erythematous base, forming circinate pattern and multiple annular lesions distributed on the posterior aspect of the lower legs. And pin head sized pustules with erythematous base recurring on the healing site on the anteriomedial side of right leg appeared(Fig. 5, 6). Results of laboratory studies showed the following values; hemoblobin, 11.3 gm/dl, hematocrit, 35%;WBC count, 7,500 cells/cu mm with 78% neutrophils, 9% lymphocytes, 1% eosinophils; ESR, 20 mm/hr; urinalysis, negative.

The value of liver function test, and electr olytes were normal. Chest X-ray showed hypertensive heart with pulmonary congestion. KOH mount was negative. Biopsy specimen of the skin was taken from the leg. In histopathological findings, there were multiple subcorneal pustules containing polymorphonuclear leukocytes with mild acanthosis in the epidermis, and diffuse chronic inflammatory

cell infiltration in the dermis, in H-E stain (Fig. 7).

She was treated with dapsone, 25 mg three times a day, orally, triamcinolone, 10 mg per ml intralesional injection, and ultraviolet light. There was a transient clinical response but complete remission was not obtained.

Discussion

On clinical grounds and on the results of investigations, it appears that patients have subcorneal pustular dermatosis. This disease first described under the name of "subcorneal pustular dermatosis" by Sneddon and Wilkinson, but Simpson[8], Carney[9] and Cipollaro[10] appear to have described patients with this disease earlier. The groin, abdomen, axillae and flexual aspects of the proximal extremities are involved frequently. Although the mucous membranes have spared in almost all cases, at least some patients have had oral lesion[11][12][13].

Subcorneal pustular dermatosis in adults has been preceded by respiratory infection in some instances and by chronic urinary tract disease in ten of 14 cases reported by Ellis. The disease has occurred in postdelivery periods, following the extract on of teeth[11], and after stopping treatment with systemically administered corticosteroids. Several observers have commented on how symptom free a patient can be before an eruption that can be so explosive and extensive occurs.

On the laboratory findings, immunofluorescent studies, direct and indirect, have not showen any abnormal fluorescence[2][3][12]. In this reported cases, immunoglobuline estimations and immunofluorescent studies were not carried out. It is emphasized that the histological appearence indicate a diagnosis of a subcorneal pustular dermatosis. Subcorneal blisters containing polymorphonuclear leukocytes occur in a number of disorders which include impetigo, subcorneal pustular dermatosis, pemphigus foliaceus and pustular psoriasis[15][20]. Someone had been considered the clinical diagnosis of pustular psoriasis; however, the histological studies showed absence of spongiform pustules, microabscess, congestion of papillae, and elongation of rete ridges that might be seen in the psoriasis.

Diagnostic confusion may occur initially between this disease and dermatitis herpetiformis, impetigo herpetiformis, and impetigo contagiosum. The more intence itching and the subepidermal location of the blister in dermatitis herpetiformis. Impetigo herpetiformis usually no itching; the blister involes the entire thickness of the epidermis; and there are profound usually systemic signs and symptoms. Impetigo contagiosum is differentiated principally on its shorter and more acute course and the findings of bacteria in the lesions, and its response to antibiotics. In reported cases, two cases have characteristic skin lesions with pruritus and typical histopathologic findings. Culture of the pustules in case 1 show abundant staphylococcus aureus probably due to secondary infection. Fungus did not detected in the skin lesion. Occasionally eosinophils may be prominent in the dermis but seldom are seen in significant numbers in vesicles. Acantholytic cells may be present near the surface of the base of blisters[20~22]. There were no eosinophilia in both reported cases.

Many forms of therapy have been tried both before and after the correct diagnosis of subcorneal pustular dermatosis has been made. It includes treatment with antibiotics; tetr acycline[23], chloramphenicol[15], thorium[24], ultraviolet light[20], tripelenamine(pyribezamine)[25], estrogen(because of premenstrual flares)[21], niacine, potassium arsenite and immunoglobu-

line. Corticosteroids administered topically, orally, and parenterally appear to be the most helpful in some instances. The drug of choice appeared to be the sulphones, such as dapsone, acetosulphone sodium(promacetin)[21], and either sulphone(diasone) sodium[26], sulfisoxazol (Gantrisin)[20][22], or sulphapyridine[1][16][20][21][27].

According to Sneddon treatment with dapsone also does not always result in complete remission[20]. However none of these drugs are uniformly successful in the treatment of subcorneal pustular dermatosis. Many physicians have observed the unusal fact that in some patients, one drug is very useful, while in others, worthless or toxic.

Treatment with vitamin E helped some patients with subcorneal pustular dermatosis[5]. In reported case 1 has been treated with combination of dapsone and corticosteroid, but complete remission was not obtained. In case 2 has been treated with combination of dapsone, ultraviolet light and triamcinolone intralesional injection.

However, in most of those patients reported, the disease is still present; with erythema multiforme[20], and impetigo; many of the patients had experienced remission and exacerbation for several years before the clinical recognition of the disease, and a benign disease with little effect on general health and longevity[15].

Summary

Two cases of subcorneal pustular dermatosis are presented. The clinical and histopathological findings are discussed.

In case 1, treatment of combination of dapsone and corticosteroids was relatively ineffective.

In case 2, treatment with combination of dapsone, ultraviolet light, and triamcinilone intralesional injection, was satisfactory clinical response transiently, but complete remission could not obtained.

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□ Explanation of Figures □

Fig. 1. Sloughing with erythematous base, forming circinate pattern on the axilla and upper arm.

Fig. 2. Confluented and discreted pustules with erythematous base on the abdomen.

Fig. 3. Stocking type of distribution of healed skin and advancing border of the active lesion of the upper thighs.

Fig. 4. Subcorneal pustules containing polymorphonuclear leukocytes.

Fig. 5. Circinate patterned sloughing and multiple annular lesions on the post. aspect of lower legs.

Fig. 6. Advancing border of the active lesion on the healing site of post. aspect of lower leg.

Fig. 7. Subcorneal pustules containing PNL with mild acanthosis, and diffuse chr. inflam. cell infiltration in the dermis.

Fig. 1.

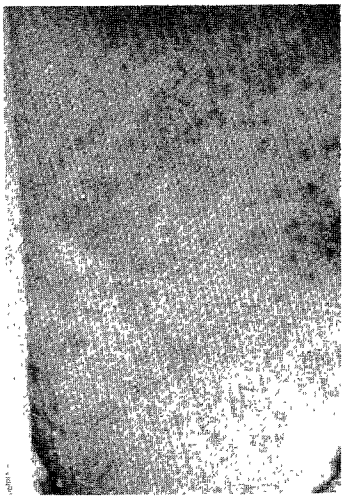

Fig. 2.

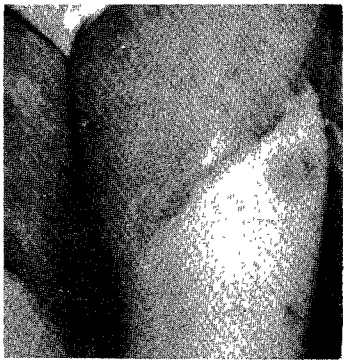

Fig. 3.

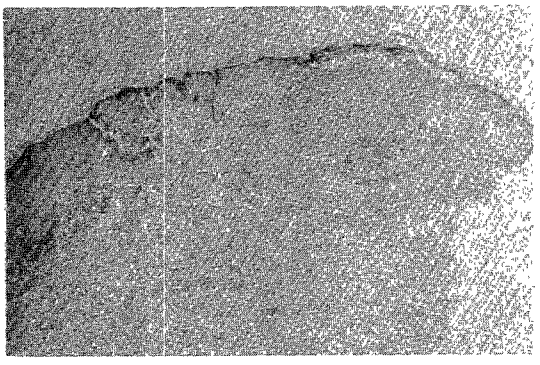

Fig. 4.

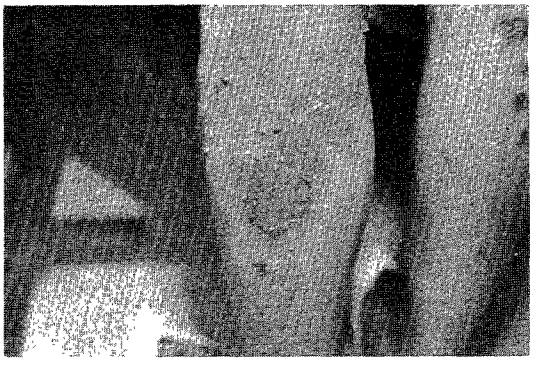

Fig. 5.

Fig. 6.

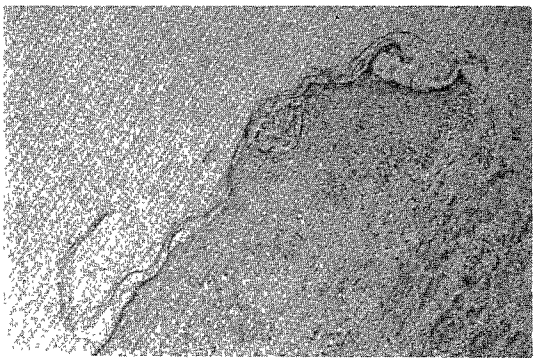

Fig. 7.