A Case of Impetigo Herpetiformis

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Impetigo herpetiformis is a rare pustular disorder that primarily affects pregnant women. A 31 year-old woman at 37 weeks and 4 days' gestation of 3rd pregnancy presented with pruritic multiple erythematous pustules and crusts on the trunk and both antecubital fossa for 3 months. She had had similar lesions in her 1st and 2nd pregnancy, which subsided after delivery without any fetal or maternal complication. She had no personal or familial history of psoriasis. Histopathologic findings showed subcorneal pustules with numerous neutrophils and spongiform pustule of Kogoj. Bacterial culture from pustules was sterile. After Caesarean section at 38 weeks' gestation, a healthy female neonate was delivered and her skin lesion resolved spontaneously. (Ann Dermatol 16(2) 83~86, 2004)

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Impetigo herpetiformis is a exceedingly rare dermatologic disease occurring particularly during pregnancy. Clinically and histologically, it bears some resemblance to pustular psoriasis. The clear similarity of these two pustular dermatoses has caused disagreement among investigators, some of whom consider impetigo herpetiformis a disease sui generis and some who include it as a variant of pustular psoriasis. It is characterized by specific cutaneous lesions and general symptoms, the disease being a serious one and not infrequently fatal. The disease recurs with every successive pregnancy with earlier and increased morbidity, and it probably is associated with increased risk of stillbirth and neonatal death[1]. We describe a case of recurrent impetigo herpetiformis which developed during 3 successive pregnancies in a 31-year-old woman, another case that supported impetigo herpetiformis and pustular psoriasis should be considered as a separate entity.

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CASE REPORT

A 31-year-old woman at 37 weeks and 4 days' gestation of 3rd pregnancy presented with pruritic multiple erythematous pustules and crusts on the trunk and both antecubital fossa for 3 months. Examination revealed multiple grouped or annular pustules and crusts on the erythematous base on the trunk, both forearms and hands (Fig. 1. A). Her skin lesions began in both sides of inguinal area and medial thigh and spread to the trunk and upper extremities. There were no constitutional symptoms such as fever, lymphadenopathy, and arthralgia. She had had similar lesions in the 3rd trimester of her 1st and 2nd pregnancy, which subsided after delivery of a healthy infant without maternal or fetal complication. There was no personal or familial history of psoriasis. Laboratory findings revealed pregnancy-related anemia of hemoglobin 9.8 mg/dl, and serum calcium and albumin levels were within normal limits. Bacterial cultures from pustules were sterile. Otherwise was nonspecific. Her current pregnancy had been uneventful with normal reactive tracing of the fetal heart monitor and the Doppler measurement showed no finding of placental insufficiency.

A skin biopsy with direct immunofluorescence was done. Histopathological findings revealed subcorneal pustules with numerous neutrophils and spongiform pustule of Kogoj. In the papillary dermis there was

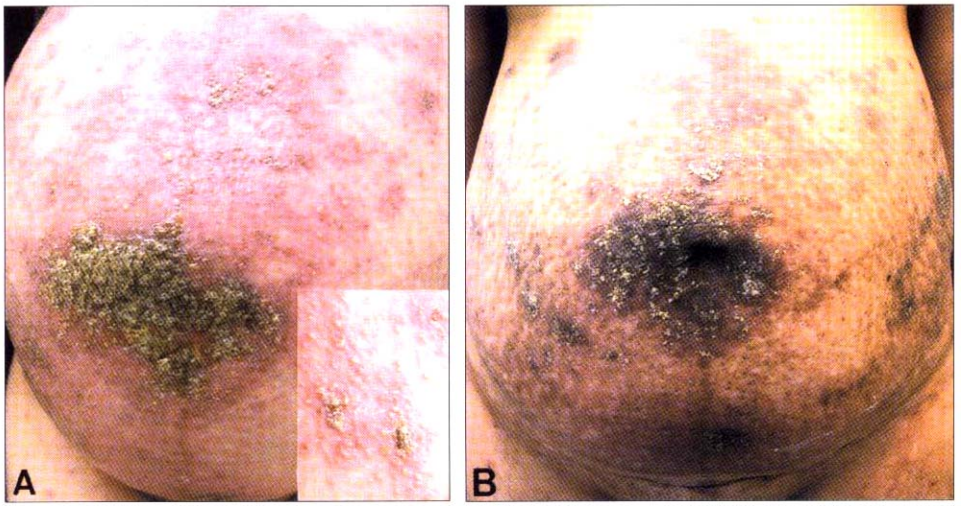

Fig. 1. A. Confluent erythematous patches studded with tiny pustules and crusts on the abdomen and its magnification (Inset). B. Marked improvement of skin lesion 10 days after Caesarean section.

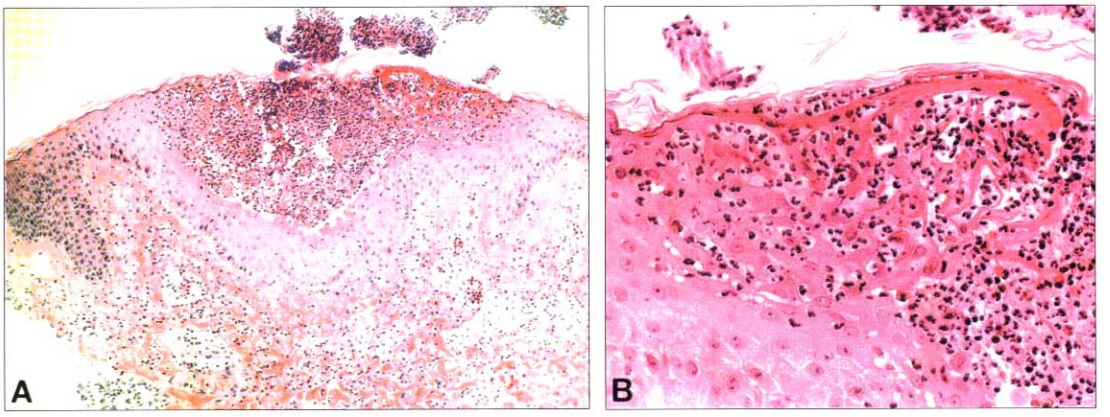

Fig. 2. A. Subcorneal pustules with numerous neutrophils and spongiform pustules of Kogoj (H & E, ×100). B. Numerous neutrophils within spongiform pustule of Kogoj (H & E, ×200).

a diffuse infiltration composed of neutrophils and lymphocytes (Fig 2. A, B). Immunofluorescence staining of the perilesional skin showed no deposition of Ig M, Ig G, Ig A or complement.

As her skin lesion was getting worse, Caesarean section was carried out at 38 weeks' gestation. A healthy female neonate (Apgar score 8/9, birth weight 3,750 gm) was delivered. After delivery, her skin markedly improved spontaneously: the erythema receded considerably, most of the pustules disappeared, and there were some crusts and postinflammatory hyperpigmentation of the affected skin areas (Fig. 1. B). After two months of follow-up period, all skin lesions resolved.

DISCUSSION

Impetigo herpetiformis was first described in 1872 by Hebra[2] as a skin disorder appearing in pregnant women and usually having a fatal outcome. This dermatosis was characterized by a disseminated spread of sterile pustules, associated with major general symptoms such as fever, diarrhea, nausea and an increased risk of abortion due to placental insufficiency. Approximately 350 cases have been re-

ported up to date in Europe and the United States1. In Korean literatures, there have been 6 cases of impetigo herpetiformis, which have showed various clinical courses[3].

Although defining the etiology of impetigo herpetiformis has been difficult due to its rarity, there is probably an etiological relationship with pregnancy. No certain pathological mechanism has been determined so far, but hypocalcemia, hypoparathyroidism, infection and oral contraceptive pills have been reported to induce impetigo herpetiformis[4-6].

Impetigo herpetiformis usually appears acutely, dramatically, and very rapidly in the latter months of pregnancy, although it may occur and has been reported to be present in the first month of pregnancy. The lesions start as marked erythematous patches occurring in the medial aspects of the thighs, groin, axillae and neck, and then spreads to other parts of the body. The typical lesions are erythematous patches that are studded with tiny superficial pustules particularly coalescing at their margins causing pain and a burning sensation. The oral and esophageal mucosae are sometimes involved. The rash is accompanied by high fever, chill, vomiting, diarrhea, lymphadenopathy and splenomegaly[3-7].

Laboratory findings include leukocytosis, an elevated erythrocyte sedimentation rate, hypocalcemia, and hyperphosphatemia. The skin lesions are sterile, and biopsy findings reveal intraepidermal pustules contiaining both neutrophils and mononuclear cells, which are the same as those of pustular psoriasis[3-7]. Our case may simulate a secondary impetigonized pruritic urticarial papules and plaques of pregnancy (PUPPP) in that skin lesions at the first visit showed pruritic, erythematous, crusted, diffuse patches mainly located on the abdomen and that there had been no general symptoms such as fever, chill and diarrhea. However, she had had similar skin lesions in her prior pregnancies and the skin lesions began with pustules primarily and coalesced to large plaques on the erythematous base, which were sterile in bacterial culture. In addition, histopathologic findings showed a typical subcorneal pustule with spongiform pustule of Kogoj. Taking all into account, we could conclude that this case of impetigo herpetiformis should be distinguished from other dermatoses of pregnancy.

The differential diagnosis includes pustular psoriasis, herpes gestationis and subcorneal pustular dermatosis. Impetigo herpetiformis bears a close resemblance to another dermatologic condition, pustular psoriasis, and it has even been suggested by some authors that impetigo herpetiformis is a variant of generalized pustular psoriasis occurring during pregnancy[4]. But others insisted that impetigo herpetiformis should be considered as a separate entity because it affects pregnant women with no previous history of psoriasis, it resolves after labors, and the patients are free from the skin disorder between pregnancies[5]. In our patient there was no past and family history of psoriasis nor attack of pustular skin lesion between pregnancies. This supports impetigo herpetiformis and pustular psoriasis should be considered as a separate entity. Herpes gestationis can be easily differentiated from impetigo herpetiformis by its clinical and histopathologic findings with subepidermal vesicles and positive direct immunofluorescent findings. Subcorneal pustular dermatosis also differs from impetigo herpetiformis in that clinically the former shows multiple annular pustules on the flexural areas and histologically there is no spongiform pustule of Kogoj. Furthermore, subcorneal pustular dermatosis doesn't have any relationship to pregnancy.

Impetigo herpetiformis tends to worsen as the pregnancy progresses. Termination of the pregnancy usually results in rapid resolution of the disease[1]. After impetigo herpetiformis has once affected a patient, it may recur in subsequent pregnancies and an earlier presentation in each subsequent pregnancy is the rule[5]. Maternal mortality has been reported, but with the use of systemic corticosteroids, the treatment of choice for impetigo herpetiformis, maternal deaths are prevented. Other serious complications of the disease is placental insufficiency and intrauterine death of the fetus. The severity and duration of the disease are the contributing factors and careful monitoring of the fetus is imperative[6].

Recommended treatment of impetigo herpetiformis includes systemic and topical corticosteroids and antibiotics for secondary infection. Systemic administration of prednisone at a dosage of 60 mg daily is necessary at times to control the eruption[1]. Once the disease is under the control, prednisone can be tapered judiciously to avoid the sudden exacerbation of the disease. The skin lesions resemble widespread burns, and that, together with the high fever, diarrhea and vomiting, result in significant fluid losses and dehydration. Fluid and electrolyte replenishment is the cornerstone of treat-

ment. Fluids and electrolytes, especially calcium and albumin, should be monitored and normalized. Maternal cardiac and renal functions should be monitored, as these can deteriorate with the progression of disease[8]. In severe cases, the pregnancy must be terminated. If the pregnancy is near term, labor may be induced; in preterm pregnancies, an attempt should be made to stimulate fetal pulmonary maturation, and then to induce labor[3-8].

We present a case of impetigo herpetiformis which began in the third trimester and showed complete resolution after delivery.

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